

**ADHERENCE TO CYSTIC FIBROSIS TREATMENT
REGIMENS: THE ASSOCIATION BETWEEN HEALTH CARE
PROVIDER RELATIONSHIP STYLES AND SELF-EFFICACY,
MOTIVATION TO ADHERE AND EMOTIONAL WELL-BEING
IN ADOLESCENTS AND YOUNG ADULTS**

BY

JACQUELINE VICTORIA SQUIRE

AUGUST 2002

**SUBMITTED IN PART FULFILMENT OF THE DEGREE OF
DOCTORATE IN CLINICAL PSYCHOLOGY AT THE
UNIVERSITY OF EDINBURGH.**



ACKNOWLEDGEMENTS

I would like to thank my Clinical Supervisor, Dr. Lesley Howells, and Dryden Badenoch for their invaluable advice, support and encouragement throughout this study. I would also like to thank my Academic Supervisor, Dr. Matthias Schwannauer, and Arthur Still, statistician, for their helpful methodological and statistical suggestions. In addition, I would like to acknowledge Dr. Somnath Mukhopadhyay and Dr. Una MacFadyen, Consultant Paediatricians, and Dr. Winter and Dr Innes, Consultant Respiratory Physicians, for their encouragement of this study and assistance in recruiting participants and also Gail Milne, Mags Pogson and Cathy Liddle, cystic fibrosis liaison nurses, for their help during the past few months, especially during the data collection. Thanks to all the adolescents and young adults with cystic fibrosis who gave their time to participate in the study and particularly to those who were willing to speak honestly and in detail about their experiences. Thanks also to my parents and sister for all their support throughout my clinical training and to my friends Nicky, Rhona and Tom, especially, for providing words of encouragement and laughs when they were much needed.

DECLARATION

“I certify that this is a true and accurate account of the work carried out. This thesis has been composed by myself with no help from others, except for those referred to in the acknowledgements”.

Signed

Jacqueline Victoria Squire

ABSTRACT

Introduction: Cystic fibrosis (CF) is a chronic condition that requires an intensive daily treatment regimen to control the symptoms and prolong life expectancy. Non-adherence to treatment, however, has been documented within the literature and is a major concern of health care professionals. Published studies investigating non-adherence have tended to neglect the role of the patient-practitioner interaction. The Self-determination Theory of Human Motivation (Deci and Ryan, 1985) highlights the importance of autonomy supportive professional relationship styles in increasing patients' self-efficacy and motivation for self-management. To date, no study has applied this theory within the CF population.

Objectives: To examine whether there is an association between health care provider relationship styles and self-efficacy and motivation to adhere in adolescents and young adults with CF. Additionally, to investigate whether emotional well-being has an effect on adherence to treatment regimens.

Design: A cross-sectional, investigative study was undertaken with CF patients, aged between 12 and 30 years, attending out-patient CF clinics in Tayside and Forth Valley.

Method: Participants completed four measures that assessed their perceptions of health care providers' relationship styles, self-efficacy, motivation to adhere and emotional well-being and provided a self-rating estimate of adherence. Semi-structured interviews were conducted with a number of participants to gather more detailed information regarding their personal experiences of CF. Demographic information was obtained from participants' hospital case notes.

Results: As hypothesised, the results suggested that autonomy supportive professional relationship styles were associated with increased motivation to adhere and self-efficacy in adolescents and young adults with CF. Patient-practitioner interactions should therefore be taken into consideration in future studies of adherence. A thematic analysis of the interview data indicated that the participants' experiences of CF were varied.

Conclusion: The research findings are discussed with reference to the current literature and clinical implications and future areas of research are identified.

CONTENTS

	PAGE NO.
Acknowledgements	i
Declaration	ii
Abstract	iii
CHAPTER 1: INTRODUCTION	1
1.0 General Introduction	2
1.1 Cystic Fibrosis	2
1.1.1 Introduction to Cystic Fibrosis	2
1.1.2 The Genetics of Cystic Fibrosis	3
1.1.3 Diagnostic Tests for Cystic Fibrosis	4
1.1.4 Life Expectancy	6
1.1.5 Clinical Features of Cystic Fibrosis and Available Treatments	7
1.1.5.1 The Respiratory System	7
1.1.5.2 Treatments Available for the Respiratory System	9
1.1.5.3 The Pancreatic System	12
1.1.5.4 Treatments Available for the Pancreatic System	13
1.1.5.5 The Gastro-Intestinal System	15
1.1.5.6 The Reproductive System	15
1.1.5.7 Other Complications	17
1.1.6 Service Delivery	17
1.1.7 Summary	18

1.2 Adherence to Treatment Regimens	19
1.2.1 Introduction to Adherence	19
1.2.2 Prevalence of Non-adherence	20
1.2.3 Consequences of Non-adherence	21
1.2.4 Measures of Adherence	21
1.2.4.1 Drug Assays	22
1.2.4.2 Observations	22
1.2.4.3 Pill Counts	23
1.2.4.4 Health Care Provider Estimates	23
1.2.4.5 Patient or Parental Reports	23
1.2.4.6 Health Outcomes	24
1.2.5 Factors Affecting Adherence	25
1.2.5.1 Patient Factors	25
1.2.5.2 Family Factors	28
1.2.5.3 Regimen Factors	29
1.2.6 Summary	30
1.3 Psychological Problems in Chronic Conditions	31
1.3.1 Prevalence of Psychological Problems in Cystic Fibrosis	31
1.3.2 The Influence of Psychological Problems on Adherence	33
1.3.3 Summary	33
1.4 Self-determination Theory of Human Motivation	35
1.4.1 Introduction to Self-determination Theory	35
1.4.2 Autonomous versus Controlled Professional Relationship Styles	35
1.4.3 Autonomous versus Controlled Motivation	36
1.4.4 Self-efficacy	37

1.4.5 Previous Research Applying Self-determination Theory	38
1.4.6 Summary	41
1.5 Present Study: Aims and Hypotheses	42
1.5.1 Aims	42
1.5.2 Hypotheses	43
CHAPTER 2: METHOD	44
2.1 Design	45
2.2 Participants	45
2.3 Measures	47
2.3.1 The Health Care Climate Questionnaire (HCCQ)	48
2.3.2 The Self-efficacy Questionnaire (SEQ)	49
2.3.3 The Treatment Self-regulation Questionnaire (TSRQ)	49
2.3.4 The Hospital Anxiety and Depression Scale (HADS)	50
2.3.5 Self-report Measure of Adherence	51
2.3.6 Semi-structured Interview	52
2.3.7 Other Information Collected	53
2.4 Procedure	54
2.5 Analysis of Data	56
2.5.1 Data Analysis	56
2.5.2 Statistical Power	57

CHAPTER 3: RESULTS	58
3.1 Demographic Data	59
3.1.1 Exploration of the Data	59
3.1.2 Participants	59
3.1.3 Summary of Demographic Data	60
3.1.4 Summary of Questionnaire Data	64
3.2 Hypotheses from the Literature	68
3.2.1 Hypothesis 1(a)	68
3.2.2 Hypothesis 1(b)	69
3.2.3 Hypothesis 1(c)	69
3.2.4 Hypothesis 1(d)	70
3.2.5 Hypothesis 2	72
3.2.6 Hypothesis 3	73
3.3 Experimental Hypotheses	74
3.3.1 Hypothesis 4	74
3.3.2 Hypothesis 5	76
3.3.3 Hypothesis 6	77
3.4 Qualitative Data	78
3.4.1 Participants	78
3.4.2 Researcher Bias	79
3.4.3 Data Analysis	79
3.4.4 Themes Identified	79
3.4.4.1 Impact of Cystic Fibrosis on Participants' Lifestyles	81
3.4.4.2 Emotional Well-being in Participants	85
3.4.4.3 Adherence Issues	89
3.4.4.4 Relationships with Health Care Providers	92

CHAPTER 4: DISCUSSION	96
4.1 General Summary of Research	97
4.2 Summary of Demographic Data	97
4.3 Discussion of the Hypotheses from the Literature	98
4.3.1 Hypotheses 1(a), (b) and (c)	98
4.3.2 Hypothesis 1(d)	102
4.3.3 Hypothesis 2	105
4.3.4 Hypothesis 3	106
4.4 Discussion of the Experimental Hypotheses	107
4.4.1 Hypothesis 4	107
4.4.2 Hypothesis 5	109
4.4.3 Hypothesis 6	110
4.5 Discussion of the Qualitative Data	113
4.5.1 Discussion of the Qualitative Findings	114
4.5.2 Methodological Issues Relating to the Qualitative Data	118
4.6 General Methodological Issues	119
4.6.1 The Recruitment of Participants	119
4.6.2 The Generalisation of the Results	121
4.6.3 The Measure of Adherence Utilised	122
4.6.4 Time Delay Incurred whilst Awaiting Ethical Approval	122
4.6.5 Post-Hoc Power Calculations	124
4.7 Clinical Implications of the Study	124
4.7.1 Implications of the Study for Health Professionals in CF Teams	124
4.7.2 Implications for Improving Adherence to Treatment Regimens	126
4.8 Future Studies	128
4.9 Conclusions	129

APPENDICES

Appendix 1: Questionnaire Pack containing:	146
(1) The Health Care Climate Questionnaire (HCCQ)	147
(2) The Self-efficacy Questionnaire (SEQ)	148
(3) The Treatment Self-regulation Questionnaire (TSRQ)	148
(4) The Hospital Anxiety and Depression Scale (HADS)	149
(5) A Self-report Measure of Adherence	150
Appendix 2: Sample Questions from the Interview	151
Appendix 3: Demographic Information Sheet	154
Appendix 4: Introductory Letter to Participants in Tayside and Forth Valley	156
Appendix 5: Information Sheet for Participants in Tayside and Forth Valley	159
Appendix 6: Consent Form	162
Appendix 7: Reply Slip for the Interview	164
Appendix 8: Letter sent to Participants' GPs	166
Appendix 9: Introductory Letter to Parents in Tayside and Forth Valley	169
Appendix 10: Information Sheet for Parents in Tayside and Forth Valley	172
Appendix 11: Reminder Letter to Participants/Parents	177
Appendix 12: Graphs of Participants' Demographic Details	182

TABLES

	PAGE NO.
Table 1: Measures used in the Current Study	47
Table 2: Summary of Patients Approached and Recruited	60
Table 3: Demographic Details of Participants	61
Table 4: Details of Responses to Questionnaire Measures	64
Table 5: Independent T-test Examining Effect of Gender on Participants' Adherence to Cystic Fibrosis Treatment Regimen Tasks	66
Table 6: Correlation Comparing Social Deprivation Category with Participants' Adherence to Cystic Fibrosis Treatment Regimen Tasks	66
Table 7: Correlation Comparing Severity of Illness with Participants' Adherence to Cystic Fibrosis Treatment Regimen Tasks	67
Table 8: Correlation Comparing Health Care Providers' Relationship Style with Participants' Motivation to Adhere	68
Table 9: Correlation Comparing Health Care Providers' Relationship Style with Participants' Self-efficacy	69
Table 10: Correlation Comparing Participants' Motivation to Adhere and Self-efficacy	70
Table 11: Correlation Comparing Participants' Self-efficacy with Adherence to Cystic Fibrosis Treatment Regimen Tasks	71
Table 12: Partial Correlation Comparing Health Care Providers' Relationship Style with Participants' Self-efficacy whilst Controlling for Participants' Autonomous Motivation to Adhere	72
Table 13: Correlation Comparing Health Care Providers' Relationship Style with Participants' Psychological Well-being	73
Table 14: Correlation Comparing Participants' Self-efficacy with their Psychological Well-being	74

Table 15: Correlation Comparing Levels of Anxiety with Adherence to Cystic Fibrosis Treatment Regimen Tasks	75
Table 16: Correlation Comparing Levels of Depression with Adherence to Cystic Fibrosis Treatment Regimen Tasks	75
Table 17: Correlation Comparing Age with Adherence to Cystic Fibrosis Treatment Regimen Tasks	76
Table 18: Correlation Comparing Severity of Illness with Participants' Psychological Well-being	77
Table 19: Descriptive Information on Participants who Attended the Semi-structured Interviews	78

FIGURES AND GRAPHS

Figure 1: The Self-determination Model	39
Figure 2: Age of Participants	62
Figure 3: Social Deprivation Categories for all Participants	63
Figure 4: Scores on the Anxiety and Depression Subscales for all Participants	65
Figure 5: The Main Categories Identified from the Interview Transcripts that Correspond to the Aims of the Study	80
Figure 6: The Self-determination Model	103
Figure 7: Diagram Depicting the Results of the Current Study	104

CHAPTER 1: INTRODUCTION

1.0 GENERAL INTRODUCTION

Cystic fibrosis (CF) is the most common life-threatening genetic disorder in the United Kingdom (Chumbley, 1999) and is estimated to affect 1 in 2500 live births in Caucasian populations (Walters, Britton and Hodson, 1993). In 1938, 70% of children with CF died within the first year of life (Andersen, 1938). However, the prognosis of the condition has improved dramatically over the years due to earlier diagnosis, the introduction of aggressive preventative medical treatment and changes in the delivery of services allowing timely interventions to be made by health care professionals. Non-adherence to CF treatment regimens has, however, been documented within the literature with serious health implications for the individual (Brown, Rowley and Helms, 1994). Recent studies have therefore become more interested in examining factors that impact on and promote adherence behaviour to treatment.

In the introduction to this study, general issues relating to CF, its treatment regimens and problems with adherence will be addressed. The influence of emotional well-being on adherence behaviours will be discussed and current literature regarding the relationship between health care providers' interpersonal styles and patient self-management will also be reviewed.

1.1 CYSTIC FIBROSIS

1.1.1 Introduction to Cystic Fibrosis

Cystic fibrosis is an autosomal recessive condition characterised by abnormal secretions of the exocrine (mucus producing) glands that lead to problems within the respiratory, pancreatic, gastro-intestinal and reproductive systems. Walters et al

(1993) estimate that there are approximately 6500 individuals within the UK presenting with the condition. Ethnic variations in the prevalence of CF are evident, however, with the prevalence of live CF births at 1 in 17,000 in African American populations and 1 in 90,000 in Oriental populations (Bush, 2001). CF requires time-consuming and demanding daily treatment regimens that aim to control the symptoms of the condition, delay further deterioration and improve the quality of life of the patient thereby helping them and their family to live as normal and productive a life as possible.

1.1.2 The Genetics of Cystic Fibrosis

CF has been recognised since the 1930's, although the CF gene was only identified in 1989 (Cuthbert, 1996). CF results from a pair of faulty genes on the long arm of chromosome 7. The gene normally produces a protein called Cystic Fibrosis Transmembrane Conductance Regulator (CFTR) that carries sodium chloride and water in and out of the cells lining the lungs and digestive tract. However, in CF the gene may fail to produce any CFTR protein, resulting in problems in the way sodium chloride and water are transported across the cell membranes and causing cell secretions to be thick and sticky (Patientwise, 1997). Current estimates claim that there are at least 200 different gene defects which give rise to the CF phenotype therefore the condition can be difficult to diagnose by genetic screening. Nevertheless, doctors and scientists are working to find ways of repairing or replacing the faulty CF gene and although gene therapy is still in the early stages, it provides hope for the future to patients and their families (Chumbley, 1999).

1.1.3 Diagnostic Tests for Cystic Fibrosis

Chumbley (1999) postulates that if a child inherits a single CF gene on one chromosome 7 and a normal gene on the other chromosome 7 then the child will be a carrier of the CF gene but will display no symptoms as the normal gene compensates. The chances of two CF carriers having a child with CF is estimated to be 25%, with a 50% chance that the offspring will be a carrier and a 25% chance that the child will not inherit any faulty CF genes.

It is possible to screen parents for the CF genotype via a DNA test on a blood sample or mouthwash and depending on the results, the health of their child can then be monitored at antenatal clinics. Following birth, a diagnostic test commonly used if CF is suspected is the sweat test. CF patients have a raised level of sodium chloride in their sweat and the sweat test measures the amount of sodium chloride in the secretions of the exocrine glands. Sweat chloride levels of greater than 60mmol are considered to be diagnostic of CF with levels between 40-60mmol considered 'borderline' and levels below 40mmol perceived as 'normal' (Shwachman, Mahmoodian and Neff, 1981). Bush (2001) also reports that measurements of sweat sodium can be helpful as sweat sodium levels are usually higher than chloride within the general population and therefore a reverse ratio in individuals suspected of having CF would be another indication of the condition. Following the sweat test, the diagnosis of CF is often confirmed by ascertaining the genotype of the patient. Cuthbert (1996) posits that the Delta F508 CFTR mutation in the gene has been detected in 70% of patients with CF in the UK.

CF may also be suspected in newborn infants if there is evidence of intestinal obstruction due to meconium ileus, the thick black substance that lines an unborn baby's gut and is passed by all newborn babies the first time their bowels are opened. In one in ten babies with CF, meconium ileus can be so thick and sticky that it blocks the bowel, causing a swollen abdomen, abdominal pain, fatty stools and vomiting. Bowel obstruction due to meconium ileus can also occur in babies without CF and therefore confirmatory sweat testing is essential according to Fakhoury, Durie, Levison and Canny (1992). Despite the diagnostic tests presently available, some children can present with poor health for months or years before CF is suspected and diagnosed. This can be physically costly as, for example, the administration of antibiotic medication to prevent or quickly treat chest infections may be delayed and subsequently the child's developing lungs could be permanently damaged. Delayed diagnosis can also create emotional difficulties within families particularly if another child with CF is born into a family without the parents having had an opportunity for genetic counselling or antenatal diagnosis.

Recent developments in postnatal screening were publicised by the Scottish Health Minister in October 2001 who announced that, from April 2002, all newborn babies in the UK would be screened for CF as part of the Guthrie heel prick test. The Guthrie heel prick test is carried out with every newborn baby aged between one and two weeks to test for various conditions such as hypothyroidism and phenylketonuria and CF was not previously included in this blood test. The inclusion of CF in postnatal screening is a significant milestone that will enable children with CF to get the best treatment at an early age, thereby extending their lifespan and enhancing their quality of life (Hill, 1998).

Around 80% of children with CF are diagnosed before four years of age having presented with diarrhoea or fatty stools, failure to thrive, poor weight gain, recurrent respiratory tract infections or a persistent cough (FitzSimmons, 1993). Variations in the severity of the disease are evident, however, with some individuals requiring few hospital admissions throughout their life whilst other individuals' health may be so poor as to necessitate regular admissions. Generally, individuals diagnosed with CF after 18 months of age are considered to have a milder form of the condition.

1.1.4 Life Expectancy

The life expectancy of individuals with CF has increased dramatically over the last thirty years due to the earlier diagnosis of the condition, advances in medical and scientific knowledge which have resulted in more effective treatments becoming available and the development of specialised centres for the management of children and adults with CF (Shale, 1996). However, it is still rare for individuals with CF to have a normal life span. In 1991, Elborn, Shale and Britton conducted an epidemiological analysis of CF patients and stated that more than 90% of children born with CF in 1990 would reach adulthood with a life expectancy of 40 years. The Cystic Fibrosis Foundation (1999) provides a more conservative estimate of life expectancy of approximately 34 years for males and 31 years for females although the reason for a poorer prognosis in females is unclear.

An increasing proportion of patients with CF are therefore surviving into adulthood. However, a consequence of this increase in life expectancy is that many of these individuals will experience additional problems to those encountered in children such as complications with diabetes and liver disease, the development of additional

infections such as *Burkholderia Cepacia* (Bush, 2001) or possible fertility problems (Shale, 1996).

1.1.5 Clinical Features of Cystic Fibrosis and Available Treatments

CF affects the respiratory, pancreatic, gastro-intestinal and reproductive systems. Problems presenting within each of these systems as a result of the condition will now be discussed in detail in addition to current treatments available.

1.1.5.1 The Respiratory System

Respiratory disease is almost universal in the CF population. Penketh, Wise, Mearns, Hodson and Balten (1987) studied 316 CF patients aged between 12 and 51 years and reported that respiratory disease was present in 99.7% of the sample. Although the lungs of CF patients are normal at birth, shortly after birth thick mucus is produced in large quantities which block the small air tubes of the lungs and can result in breathing difficulties and a constant cough. The thick mucus within the lungs is believed to be the primary reason for the recurrence of chronic respiratory infections. The mucus can become infected with a range of viruses that allow bacteria and certain fungi to colonize within the respiratory tract leading to secondary infection that causes production of additional mucus. Individuals with CF also produce only small quantities of natural anti-bacterial chemicals within their bodies and are therefore prone to persistent respiratory infections which can cause lesions in the lung tissue over time and may ultimately lead to respiratory or heart failure. Consequently, lung function tests are regularly conducted at CF clinics to ascertain how well the lungs are functioning.

Cough swabs or sputum samples are also taken within CF clinics during infections to determine the bacteria present. The most feared broncho-pulmonary infection for individuals with CF is *Burkholderia cepacia* which is resistant to virtually every antibiotic and may cause a septicaemic illness in CF patients previously doing relatively well (Lewin, Byard and Davis, 1990). In 1998, 3.5% of American CF patients were known to be colonized with *Burkholderia cepacia* (Cystic Fibrosis Foundation, 1999). Allergic broncho-pulmonary aspergillosis, characterised by the plugging of the lobar and segmental bronchi with mucus, is also reported to affect approximately 15% of CF patients and is a difficult fungus to treat (Shale, 1996). In addition, *Pseudomonas aeruginosa* is becoming increasingly problematic within the CF population, especially in individuals who have received multiple courses of antibiotics. Although *Pseudomonas aeruginosa* is considered to be less infectious than *Burkholderia cepacia*, Shale estimates that over 80% adults with CF will eventually become colonized with the organism. Early, aggressive treatment delays colonization but once present, eradication of the organism is not possible (Valerius, Koch and Hoiby, 1991). Consequently, the initial colonization of *Pseudomonas aeruginosa* is often seen as a watershed by patients and their families (Bush, 2001). CF patients presenting with *Burkholderia cepacia* or *Pseudomonas aeruginosa* are now segregated from other CF patients during hospital admissions and clinic appointments to reduce the risk of spreading the bacteria. This action, however, can negatively impact on the social networks of infected individuals (Smith, Smith and Gumery, 1992).

1.1.5.2 Treatments Available for the Respiratory System

Current treatment for the respiratory system aims to keep the airways clear of excessive secretions and cure infections quickly and effectively to prevent further pulmonary deterioration.

Chest Physiotherapy

Chest physiotherapy has been described as the “cornerstone of treatment” for respiratory disease (Chumbley, 1999). Physiotherapy techniques involve postural drainage, percussion and directed coughing (Shale, 1996). These methods help to dislodge the thick, sticky mucus from the lungs and enable the individual to expectorate the secretions from the lungs. Physiotherapy also helps to prevent pulmonary infection by loosening and removing the mucus secretions that obstruct the airways and provide an environment for bacteria and viruses. The amount of physiotherapy required depends on the individual's health but generally physiotherapy is undertaken twice a day for ten to fifteen minutes, and even longer if the individual has a chest infection since more mucus is produced. In addition to physiotherapy, breathing techniques help to relax the airways and clear secretions and the use of a PEP (Positive Expiratory Pressure) mask can assist patients to achieve smooth and steady breathing patterns. Although parents are usually responsible for administering physiotherapy in childhood, other family members and friends are encouraged to learn the techniques so that the child is not solely dependent on their parents. Physiotherapy is time-consuming and the incorporation of the treatment into the child's daily routine from an early age is recommended to encourage adherence.

Medication

Some CF centres favour preventative medical treatment and commence children on continuous oral antibiotics from infancy. Furthermore, they may advocate regular three monthly courses of antibiotics regardless of the patient's current clinical state, especially if the individual is colonized with *Pseudomonas aeruginosa* (Szaff, Hoiby and Flensburg, 1983). Other CF centres, however, prefer to introduce antibiotic treatment during times of pulmonary infections to relieve the symptoms and reduce inflammation. Antibiotics can be taken orally, through a nebulizer machine or via an intravenous drip. Nebulized antibiotics are usually administered following physiotherapy when the lungs have less mucus in them and the machine converts the liquid medication into a fine mist which is inhaled directly into the lungs. If an individual has a serious respiratory infection they may be admitted to hospital for a course of antibiotics lasting a fortnight. Moreover, if frequent intravenous antibiotics are required or there are problems with venous access, a port-a-cath can be inserted into the patient's chest wall so that treatment can be administered at home or in hospital, as and when required, without having to administer an injection every time. Although port-a-caths are unobtrusive, adolescents in particular may be reluctant to have one inserted due to concerns about body appearance. Furthermore, intravenous antibiotics can be inconvenient to administer and are an additional burden to families who already engage in time-consuming daily treatment regimens. Families are therefore carefully assessed prior to deciding whether intravenous treatment should be administered at home or in hospital. Nevertheless, enabling some individuals to administer their intravenous antibiotics at home prevents them from spending long periods away from their family as these can be difficult for all parties and can also be disruptive to the patient's schooling or employment.

In addition to antibiotic medication, many patients with CF take the enzyme DNase through a nebulizer before physiotherapy to aid expectoration. DNase became available in 1995 and breaks down and dilutes the mucus in the airways, making the secretions more free-flowing and easier to clear by physiotherapy or directed coughing. Bronchodilators can also be prescribed for use in a nebulizer machine prior to physiotherapy, to aid sputum clearance by opening the airways.

A third of children with CF also present with asthmatic symptoms and require medication such as corticosteroids which can be administered by either an inhaler or a nebulizer. Corticosteroids reduce the inflammation in the airways and subsequently reduce wheezing and relieve shortness of breath. However, Shale (1996) reports that the continued use of inhaled corticosteroids can have considerable side effects such as growth failure and the precipitation or deterioration of diabetes.

Exercise

Regular exercise is encouraged to provide a good supply of oxygen to the lungs to enhance the individuals' ability to fight infections and help clear the sputum from the lungs. A reduced exercise tolerance can also provide an indication of a deterioration in the individual's lung function.

Lung Transplantation

Respiratory failure is the leading cause of morbidity and mortality in the CF population and is responsible for 97% of all CF deaths and 75% of all hospital admissions (Penketh et al, 1987). Lung function is commonly measured at CF clinics by forced expiratory volume at one second (FEV1). Each CF patient is assigned a

predicted FEV1 based on their gender, age and height and the FEV1 % predicted gives an indication of the capacity to which the individual is functioning. Lung transplantation will be suggested if a CF patient presents with a severe deterioration in lung function, as indicated by a FEV1 of less than 30% of their predicted value, requires increased hospitalisations, displays progressive weight loss and an impaired quality of life (Dark and Corris, 1996). Bilateral lung transplantations are usually undertaken due to the risk of spill-over infection if only one lung is replaced. Dark and Corris report that the suggestion that a patient requires a lung transplant can be a highly stressful time for both patients and their families as it implies that there is a serious risk of death due to respiratory failure. However, although complications can arise post transplant, such as organ rejection and upper respiratory tract infections, lung transplantation can be highly beneficial to patients with a 70-75% survival rate one year after transplantation and a significant improvement in their quality of life (Dark and Corris, 1996).

1.1.5.3 The Pancreatic System

Abnormally thick and sticky mucus is also developed within the pancreas in 89% of individuals with CF resulting in pancreatic insufficiency (Penketh et al, 1987). The pancreas is the digestive organ that lies behind the stomach and produces chemicals called insulin and glucagons that control the amount of sugar in the blood and produce digestive juices (pancreatic enzymes) that pass into the small intestine and break down the fats, proteins and carbohydrates consumed so that they are absorbed. In CF, the mucus produced obstructs the pancreas and hinders the digestive juices from reaching the intestines to help break down and digest food, thereby preventing the food from being properly absorbed. The mucus also prevents the digestive juices

from neutralising the stomach acid leading to the development of cysts and fibrosis (scarring and thickening) in the stomach. Pancreatic insufficiency is often recognised by abdominal distension, abdominal pain, fatty and offensive smelling stools, a failure to gain weight and appetite loss.

1.1.5.4 Treatments Available for the Pancreatic System

Pancreatic Enzymes

Due to pancreatic insufficiency, the majority of individuals with CF are required to take pancreatic enzymes, such as Pancreatin or Creon, throughout their life with every meal and snack to replace the missing enzymes and counter the digestive problems and malnutrition that would otherwise result (Shale, 1996). Pancreatic enzymes are administered in the form of medication, powder or capsules and are released in the intestine to block the gastric acid secretion that reduces enzyme activity and thereby to accelerate the breakdown of food for absorption. Individuals are required to titrate the dose according to the size of the meal or snack they consume and their faecal fat measurements. The correct dose can often take time to establish although problems with constipation tend to indicate that too many enzymes have been consumed. If pancreatic enzymes are missed regularly then digestion is affected and the individual will be at risk of nutritional deficiency, will fail to gain weight and may become lethargic. Poor weight gain and low energy levels can, however, also result from the increased resting energy expenditure and increased breathing related to an infection and subsequent deterioration in respiratory function.

High Calorie Diet

Due to the problems associated with the pancreas, individuals with CF are encouraged to sustain a diet that is high in calories, fibre and fat with a recommended calorie intake of 120-150% of the usual daily allowance, although additional calories are required if the individual is unwell. Regular meals and snacks are also encouraged with the aim of maintaining the individual's height and weight within the normal range and preventing malnutrition and physical weakness (Nasr, 2000). Improved nutrition can also benefit immunological status as fat is the most concentrated source of energy and energy and protein intake need to be high to fight infections (Bush, 2001). Better nutrition therefore results in fewer chest infections and a slower deterioration of pulmonary function, thereby improving the chance of surviving into adulthood (Cuthbert, 1996). If a CF patient exhibits extreme weight loss they may require tube feeding to ensure a high-energy intake and improve their weight gain. Tube feeding is often undertaken at night via a nasogastric tube or gastrostomy directly into the stomach with the tube attached to a bag of calorie and nutrient-enriched food. Feeding supplements such as Fortisip may also be recommended.

Vitamin, Salt and Calcium Supplements

Vitamin deficiencies are common in patients with CF, particularly the fat-soluble vitamins A, D, E and K (Shale, 1996). Due to the malabsorption of these vitamins from foods consumed, vitamin supplements are usually required. Salt supplements may also be necessary, especially in hot weather, because the significant amount of salt lost in the individual's sweat puts them at risk of salt depletion, dehydration and heat stroke. Furthermore, calcium supplements may be required due to a lack of fat-

soluble minerals such as zinc as this can have a negative effect on the bones and put the individual at risk of osteoporosis.

1.1.5.5 The Gastro-Intestinal System

One of the most common presentations of CF in newly born infants is meconium ileus which, as mentioned earlier, is the substance that lines an unborn baby's gut and is passed the first time their bowels are opened. However, meconium ileus can be so thick and sticky in CF patients that it not only blocks the bowel but can lead to a perforation of the gut. In older CF patients, the problem is generally referred to as meconium ileus equivalent or distal intestinal obstruction syndrome and causes temporary food intolerance, abdominal pain and constipation. In their report of 316 CF patients, Penketh et al (1987) claimed that acute meconium ileus equivalent was evident in 16% of their sample and that a partial obstruction of the bowel was evident in an additional 19% of the sample studied. Bush (2001) also asserts that individuals with CF may have problems with an intestinal obstruction in later life due to poorly digested food. In addition, CF patients may present with rectal prolapse, weakened by the amount of bulky stools passed or the severe bouts of coughing experienced due to respiratory infections. Treatment to alleviate intestinal obstructions usually requires an enema or oral lactulose although surgery may be necessary in acute cases to relieve the blockage or remove the blocked part of the bowel. Surgery may also be required to relieve problems associated with a prolapse of the rectum.

1.1.5.6 The Reproductive System

Growth and puberty are delayed by about two years in individuals with CF although sexual development is otherwise normal in both sexes (Reiter, Stern and Root, 1981).

Female patients may present with a delayed menstrual cycle or amenorrhoea, partly due to their low weight and partly due to the lack of nutrients within the body which causes the body's systems to shut down to preserve the nutrients for those areas that need them more urgently. Females with CF may therefore experience difficulty conceiving due to this delay in menarche and also the mucus in their cervix that can act as a barrier to sperm. Although many do conceive naturally or through assisted conception and thereafter successfully carry their pregnancy to term, the physiological stress of pregnancy and childbirth can put significant strain on the female's cardio-pulmonary functioning (Edenborough, Stableforth, Webb, Mackenzie and Smith, 1995). Generally clinical practice for medical staff is to advise against pregnancy in women with poor respiratory function and low weight as they may not gain adequate weight during pregnancy and could be at risk of death during the stress of labour. With regard to male CF patients, Gotz and Gotz (2001) state that infertility is almost universal due to the obstruction of the vas deferens with mucus. Brugman and Tausig (1984) estimate that approximately 98% of males with CF are infertile although state that in vitro fertilisation is possible in some cases if enough sperm is obtained. Reproduction issues can place additional strain on adolescents and young adults with CF compounding developmental changes already in existence at this time of life. Furthermore, because these issues could subsequently have an effect on the individual's ability to form and maintain relationships and impact on their psycho-sexual functioning, further research into this area may be important.

1.1.5.7 Other Complications

Diabetes Mellitus

According to Shale (1996), the development of CF related diabetes during adolescence or later life is a common complication for 30-50% of individuals with CF because of pancreatic dysfunction which causes a deficiency in insulin and results in abnormally high levels of sugar in the blood. CF related diabetes differs from either Type 1 (insulin-dependent) or Type 2 (non-insulin dependent) diabetes as it shares features of both types of diabetes (Nasr, 2000). The development of diabetes can be an additional burden for CF patients as they have to contend with further treatment regimens to those already required for their CF, such as regular glucose testing and insulin injections. Nevertheless, the individual must continue with the high calorie diet required for their CF condition as opposed to the standard diabetic diet.

Liver Disease

There is evidence that mild liver disease is present in 29% of the adult CF population (Penketh et al, 1987) and that the prevalence of this hepatobiliary complication increases with age (Roy, 1983).

1.1.6 Service Delivery

Service delivery has developed in time with specialist CF clinics being created for the management of children and adults with CF. Warwick (1982) states that treatment appears to be most effective and life expectancy enhanced when health care is delivered by a specialist team familiar with the patient. CF teams tend to be multi-disciplinary in nature and include consultant respiratory physicians, CF liaison

nurses, dieticians, physiotherapists and, occasionally, clinical psychologists and pharmacists. This skill mix is considered to be an additional strength of specialist teams. Clinics are generally held on a monthly basis and provide patients, and their families where appropriate, with an opportunity to meet with each professional to discuss issues relating to their health and current treatments. The CF liaison nurses tend to meet with patients at their homes in between attending to them at the clinics for routine check ups. The transfer from paediatric to adult CF clinics usually occurs between the ages of 16 to 18 years (Westwood, Henley and Willcox, 1999). Nasr (2000) states that this transition should be undertaken gradually and involve the active participation of the patient, their family, the paediatrician and adult health care providers. Rosen (1995) agrees and claims that the transfer to adult care is more likely to be successful if it is well timed, is preceded by adequate groundwork and is individually tailored for each patient. Support should also be made available for parents at this time to allow them to voice any concerns about the transfer of responsibility for the daily management of the condition to their child.

1.1.7 Summary

Individuals diagnosed with CF and their families have to learn to live with the uncertainty of a chronic disease with an unpredictable prognosis and adapt their lifestyles to incorporate intensive treatment regimens to maintain as normal a life as possible. Improvements in the diagnosis of the condition and the early introduction of treatments have resulted in prolonging the life expectancy of CF patients. Consequently, it is likely that service provision for this population will require continual expansion and greater attention will need to be focused on the physical and psycho-social course of this disease from childhood into adulthood.

1.2 ADHERENCE TO TREATMENT REGIMENS

1.2.1 Introduction to Adherence

Within the health care setting, treatment recommendations to improve the health outcome of patients are made on the assumption that they will be adhered to. Many studies, however, have demonstrated that adherence to treatment regimens is less than optimal and clinicians often have difficulty in understanding why people do not adhere to prescribed regimens, particularly in chronic health conditions (D'Angelo and Lask, 2001). Conway, Pond, Hamnett and Watson (1996) advocate that it is important to investigate the extent of non-adherence in health conditions and patient characteristics associated with incomplete adherence so that these factors can be considered when appraising a patient's condition and planning future treatment programmes. By understanding the reasons why patients do or do not adhere to treatment regimens, strategies can then be developed and applied by health care professionals to promote self-care.

Adherence has been defined as "the extent to which a person's behaviour (in terms of taking medications, following diets or executing lifestyle changes) coincides with medical or health advice" (Haynes, 1979. p1-2). Rapoff (1999) asserts that this definition is one of the most widely quoted in the literature as it focuses on specific behaviours required in prescribed treatment regimens and does not view adherence as an 'all or nothing' phenomenon but instead considers the concordance between what the patient is being asked to do and what they actually do. The term 'compliance' was utilised in early studies on adherence but was thought to imply an authoritarian approach to health care, requiring unquestioned obedience by patients to health care

providers' recommendations, rather than an active participant in the treatment process (DiMatteo and DiNicola, 1982). Consequently, the term 'adherence' is now more frequently applied within the literature and this reflects an important shift in thinking about the delivery of health care. Rapoff postulates that effective health care requires a co-operative relationship between patients and providers, where the patient feels understood and able to discuss treatment difficulties with their provider, since it is the patient who determines whether or not they will engage in the prescribed regimen to lessen the impact of an illness.

1.2.2 Prevalence of Non-adherence

The prevalence of non-adherence to treatment regimens appears to vary depending on the health condition, the age of the patient, the regimen required, the measure of adherence applied and the criteria used to classify acceptable levels of adherence. Studies have suggested that non-adherence is particularly problematic in chronic health conditions. Rapoff and Barnard (1991) estimate that about a third of patients do not adequately adhere to regimens for acute conditions whilst between 50-55% do not adhere to regimens for chronic conditions. Conway et al (1996) also claim that patients with a chronic disease only comply with about 50% of their treatment and suggest that adherence may be less than optimal since chronic health conditions often require more complex and time-consuming daily regimens.

Passero, Remor and Salomon (1981) measured adherence to CF treatment regimens via out-patient self-report. The regimens included antibiotics, vitamins, chest physiotherapy and diet and Passero et al concluded that whilst 90% of out-patients were completely adherent to antibiotics and vitamins, only 40% complied fully with

chest physiotherapy and 20% followed dietary recommendations. Czajkowski and Koocher (1987) described an adherence rate of 65% to prescribed treatment regimens, including chest physiotherapy and medication, in hospitalised adolescents and young adults with CF, although they also found that adherence varied across treatment components.

1.2.3 Consequences of Non-adherence

Non-adherence to treatment regimens can have significant consequences for the health outcomes and quality of life of patients (Brown et al, 1994). Within the CF population, non-adherence to antibiotic medication is linked to the re-emergence of infectious diseases that have become resistant to previously effective antibiotic drugs and could be particularly threatening to the well-being of the individual due to their reduced immunity. Furthermore, physicians may attribute poor outcome to inadequate treatment regimens and prescribe additional medication or they may attribute treatment failures to non-adherence and fail to make necessary changes to the regimen (Rapoff, 1999). Non-adherence to regimens also has financial consequences in terms of the money spent on unused medication and the cost of unnecessary increases in hospital admissions due to the resultant deterioration in health.

1.2.4 Measures of Adherence

A variety of techniques are available to measure levels of adherence including drug assays, observations, pill counts, health care provider estimates, patient or parental reports and health outcomes. There is, however, no ideal measurement of adherence at present with all available techniques having at least some methodological

limitations (Rapoff, 1999). The advantages and disadvantages of each of the aforementioned techniques will now be discussed.

1.2.4.1 Drug Assays

Assays are useful to confirm that medication has been ingested and to determine sub-therapeutic, therapeutic and toxic levels of drugs. Drug assays also do not rely on potentially inaccurate estimates of adherence from patients, parents or health care providers. However, assays measure adherence over relatively short time intervals and therefore fail to provide information about the consistency of adherence over extended periods of time (Rapoff, 1999). Furthermore, drug assays may be affected by differences in the way drugs are absorbed, metabolised and excreted and results may therefore be unrelated to adherence. Assays are also expensive and invasive and are therefore less clinically feasible.

1.2.4.2 Observations

Observations can be used to evaluate patient techniques in performing skills necessary for adherence, such as insulin administration for CF related diabetes. Observation methods enable clinicians and researchers to measure the frequency and duration of particular behaviours and avoid possibly misleading subjective judgements about adherence behaviours. However, observations do not provide information on how often or consistently an individual performs particular behaviours when they are not being observed. There is also the risk that patients may behave differently if they are aware that they are being observed, for example, they may be more adherent. Moreover, accessibility to adherence behaviours can be

problematic. Observations are also labour-intensive and variations between observers in recording behaviour over time may be evident.

1.2.4.3 Pill Counts

This measure of adherence involves counting medication at two points in time separated by, for example, a one-week period. Pill counts can be conducted in clinical settings or information can be provided by patients or parents. However, pill counts do not confirm ingestion of the medication and the information obtained does not provide an indication of variations in drug administration over time, such as overdosing. Furthermore, some researchers have suggested that pill counts should not be utilised to measure adherence due to significant evidence that they overestimate adherence behaviour (Bond and Hussar, 1991).

1.2.4.4 Health Care Provider Estimates

Health care provider estimates involve ratings by physicians or nurses of the degree to which they perceive their patient to be adherent to particular regimen components, often through the use of a Likert rating scale. Provider estimates are simple and inexpensive and thereby feasible to use in clinical settings. Nevertheless, Rapoff (1999) maintains that although provider estimates are generally accurate in identifying adherent patients, they may fail to identify non-adherent patients.

1.2.4.5 Patient or Parental Reports

Patient or parental reports of levels of adherence are usually obtained via rating scales, structured interviews or diary keeping and are convenient, inexpensive and clinically feasible (Bond and Husser, 1991). Patient or parental estimates can,

however, be sensitive to social desirability effects, whereby patients or families tell providers what they believe they want to hear, and reports may therefore tend to overestimate adherence by minimising non-adherence. In order to reduce the likelihood of socially desirable responses, Rapoff (1999) states that non-judgemental questions about adherence should be utilised as they are less likely to create defensive reactions in the respondent, thereby increasing the likelihood that more accurate information regarding adherence levels will be provided. Rapoff also reports that patients or parents are often asked to rate adherence over unspecified time intervals and comments that their estimates may be subject to recall errors. Rudd (1993) suggests that the “outer limits” for recall for events is generally less than two weeks and recommends that estimates of adherence should be obtained within a time period of less than two weeks. Subjective reports are preferable to parental reports since parental estimates of adherence are not considered to be as accurate since parents may not always have access to relevant adherence behaviours, especially during adolescence, and can therefore only report on what they see thereby possibly underestimating levels of adherence.

1.2.4.6 Health Outcomes

Physiological outcome measures such as pulmonary function tests in CF are useful in examining the clinical utility of behaviour changes and determining whether adherence is sufficiently problematic to require intervention (Dunbar and Agras, 1980). However, Rapoff (1999) states that health outcome measures should not be used as sole indicators of adherence since physical health is multiply determined.

The aforementioned measures of adherence differ in terms of cost and feasibility for use in clinical settings. Patient or parental reports regarding adherence remain the most direct and practical way of assessing adherence in clinical practice as patients and their family members have a unique perspective on how a chronic condition and its treatment affect their lives (Rapoff, 1999). Masek and Jankel (1982) suggest that the most effective measure of adherence is achieved by combining a variety of techniques and Rapoff posits that regular measures of adherence should be obtained to provide a more representative view of actual adherence to treatment regimens. In addition, Lask (1994) postulates that adherence behaviour should be conceptualised along a continuum as opposed to individuals being defined as either ‘adherent’ or ‘non-adherent’.

1.2.5 Factors affecting Adherence

It is important to examine predictors of adherence behaviour since any negative correlates consistently identified in studies can be utilised to develop ‘risk profiles’ which health care providers can use to identify those patients likely to present with difficulties in adherence to regimens (Rapoff, 1999). Attempts can then be made to modify those predictors related to non-adherence. To date, research has focused on investigations of specific patient, family and regimen factors that influence non-adherence and these factors will now be discussed.

1.2.5.1 Patient Factors

Studies by Lorenz, Christensen and Pichert (1985) and Patterson (1985) have found that boys are less adherent than girls to regimens for diabetes and CF respectively and Patterson has also suggested that lower socio-economic status is associated with

non-adherence to treatment regimens within these two medical conditions. The age of the patient can also significantly affect adherence to treatment regimens with adherence during adolescence and early adulthood being considerably more problematic than in younger children and this will be the main focus of this section.

Within CF, parents, especially mothers, often have the major responsibility for the daily management of their child's condition and studies by Gudas, Koocher and Wyplj (1991), Bond, Aiken and Somerville (1992) and Patterson (1985) have reported that adherence to chronic conditions such as CF and diabetes is generally better in childhood than during adolescence when parents begin to transfer responsibility for self-care to the adolescent.

Adolescence is renowned as a time of major physical, cognitive, emotional and social change when the development of a sense of independence and the establishment of peer relationships becomes increasingly important (Tyrrell, 2001). Conway (1998) reports that adolescence can be a challenging and turbulent time due to the confusing and conflicting emotions being experienced. The presence of a chronic illness such as CF can, however, magnify the normal problems associated with this age group since adolescents with CF also have to deal with assuming greater responsibility for their condition and its self-management from their parents as parental assistance decreases and cope with the transition from paediatric to adult services. Adherence in adolescents with CF is therefore considered to be a more complex issue.

Adolescence is also regarded as a time of experimentation and risk taking behaviour and many adolescents commonly reject routines and authority in their search for independence (Tyrrell, 2001). The implications of teenage rebellion can be serious, however, if the adolescent has a chronic condition. Adolescents with CF may rebel against the burden of their treatment and neglect regimen tasks, such as physiotherapy, due to their time-consuming nature which can conflict with after school activities. Parental reminders to adhere may also be perceived by the adolescent as controlling and may enhance their resolve not to adhere. In addition to resulting in conflict with their parent or health care provider, refusal to conduct CF treatment programmes can put the adolescent at risk of developing health complications in later life (Pownceby, 1997). Desmond, Schwenk, Thomas, Beaudry and Coates (1983) stated that after missing three weeks of physiotherapy a measurable lung function deterioration can be detected although there may not be an immediate effect on the individual's health.

The desire for social acceptance by peers at this time can also outweigh the demands of a self-care regimen (Cameron and Gregor, 1987). Within the school environment, adolescents may be resistant to undertake aspects of their treatment such as taking pancreatic enzymes at mealtimes due to their embarrassment at being asked the purpose of the tablets. Having to take enzymes with every meal and snack can also emphasise that the individual is different from their peers and consequently adolescents may decide to take the enzymes in secret or may not take them at all. Adherence to pancreatic enzymes can therefore become problematic at school. However, good nutrition is of major importance to the long-term survival of CF patients (Corey, McLaughlin, Williams and Levison, 1988) and therefore adherence

to the dietary aspects of the regimen is vital. Unfortunately, the focus on weight and body image and peer pressure to diet during adolescence can also result in reduced adherence to these aspects in order to remain at the same weight. The consequences of weight loss for the health of adolescents with CF can be severe and irreversible and the development of anorexia nervosa, accompanied by a significant deterioration in the individual's health, is a real risk.

In relation to young adults with CF, Duncan-Skingle and Pankhurst (2001) reported that many young adults try to identify with their healthy peers as they move towards early adulthood and brush aside the reality that they have a progressive life-threatening illness in order to try to enjoy their life. Shale (1996) states that it is commonplace for young adults to ignore the signs and symptoms that point to a deterioration in health which can be problematic particularly since many young adults with CF die within the second or third decade of their life. Gudas et al (1991) also found that young adults adhered less to medication and hypothesised that as their disease becomes more severe with age, the amount of medication required increases. Consequently, the treatment regimen becomes more intrusive for the young adults and provides a continuous unwanted reminder of the presence of the disease.

1.2.5.2 Family Factors

As children approach adolescence, parents and health care professionals are faced with the difficult task of remaining in a supportive role whilst encouraging the adolescent to assume greater responsibility for treatment and become more independent. Adherence has been found to be poorer when there is parental-child

conflict or ambiguity amongst family members regarding who has primary responsibility for undertaking the regimen tasks. A lack of parental monitoring and supervision during adolescence has also been found to contribute to non-adherence and it is emphasised that adolescents, in particular, require support and assistance from their family to maintain emotional and physical stability during this developmental stage (Conway, 1998). Adolescents and young adults are also more likely to adhere with treatment regimens if they have a close relationship with family members and friends and come from an organised family life where a routine has been established to undertake treatment, whilst permitting the individual to lead as normal a life as possible.

1.2.5.3 Regimen Factors

With regard to regimen factors, Gudas et al (1991) assert that patients who are less knowledgeable about their condition and its treatment are less adherent to regimens. Haynes (1979) stated that the complexity of the regimen, the duration and severity of the illness and the intrusiveness of the treatment regimen were related to poorer adherence. Families have a finite amount of time, energy and resources to devote to medical regimens particularly if they are to try to maintain some semblance of a 'normal' family life (Patterson, 1985). As would be expected therefore, Glasgow, McCaul and Schafer (1986) and Passero et al (1981) have stated that adherence tends to be lower with more complex regimens such as chest physiotherapy in CF and dietary regimens for diabetes. The perceived benefit of the regimen tasks has also been associated with inconsistent adherence. D'Angelo and Lask (2001) report that adherence in CF is better when the regimen task is considered to have an immediate effect on the health of the individual, for example, severe abdominal pain if

pancreatic enzyme replacements are not consumed with every meal and snack. Treatments such as physiotherapy are often perceived by patients and their families as not having any immediate benefits and may therefore not be regularly undertaken. Physiotherapy in particular is also time-consuming and impinges on daily activities (Pownceby, 1997). It is therefore important to provide information on the efficacy of regimens to demonstrate that patients and their family members' efforts to maintain optimal adherence are having the desired beneficial effect. Furthermore, since variations in adherence to different components of treatment have been reported, Passero et al (1981) state that it is important to consider all aspects of regimen tasks equally when assessing adherence behaviour.

1.2.6 Summary

Non-adherence to treatment regimens is a commonly acknowledged problem in health care settings in both adolescents and young adults with CF and reasons for non-adherence appear to be multiply-determined. There are significant consequences for the health outcomes of patients who do not adhere and Rapoff (1999) posits that efforts to change adherence behaviour should be considered when non-adherence is evidently compromising the health and well-being of the patient. In view of the significant problems with non-adherence reported within young people with CF it is considered an important area for studies to investigate with the aim of developing strategies to encourage improved adherence. The impact of psychological problems on adherence will be discussed within the next section of this chapter and the influence of the practitioner-patient interaction on adherence to treatment regimens will be discussed in section 1.4.

1.3 PSYCHOLOGICAL PROBLEMS IN CHRONIC CONDITIONS

1.3.1 Prevalence of Psychological Problems in Cystic Fibrosis

Research has been undertaken to examine the prevalence of psychological problems within chronic conditions. Cappeli et al (1987) reported that living with a chronic disease can be a considerable source of stress for children and adolescents due to the limitations to their freedom. Furthermore, Kyngas and Barlow (1995) interviewed 51 adolescents aged between 13 and 17 years with insulin dependent diabetes mellitus to determine the impact of diabetes on their lives. The results suggested that diabetes had an effect on the psychological well-being of participants and was associated with fear, depression, worry, stress and guilt. In particular, participants presented with anxiety about their ability to cope with possible future complications and depression about the limitations imposed by the need for regular, life long insulin treatment. Feelings of guilt were also reported if the participant had been neglecting their self-care. Kyngas and Barlow concluded that having a chronic disease was perceived by the adolescents as a threat to their psychological, social and physical well-being.

Within the CF population, early research also suggested a high incidence of emotional disturbance, particularly depression, in children (Lawler, Nakielny and Wright, 1996). In addition, Aspin (1991) reported that CF patients were prone to develop symptoms of anxiety or depression as they had little sense of control over their illness and Thompson, Hodges and Hamlett (1990) reported a high incidence of anxiety. Cowen et al (1984) also investigated the prevalence of psychological distress in 191 CF patients aged 16 years and above and found that participants presented with significant levels of psychological distress. In particular, the results suggested that 12% of males and 30% of females with CF aged between 16 and 19

years displayed moderate or severe levels of emotional distress whilst the frequency of distress doubled in both males and females over the age of 20 years. Moreover, Strauss and Wellisch (1981) reported that 43% of their sample of adult CF patients reported feeling depressed 'occasionally' or 'frequently' and claimed that this was due to their concerns regarding their physical appearance, the constant treatments required for the condition and uncertainty about their future health. Adults with CF are required to undertake similar treatments to adolescents in order to maintain body weight, nutrition and lung function (Shale, 1996). However, adults also have to deal with additional issues such as the development of liver disease and diabetes, male infertility, concerns regarding pregnancy, issues relating to career prospects and independent living and the prospect of limited survival. These additional factors may account for the reports of increased psychological problems within this age group.

Nevertheless, despite studies suggesting high rates of psychological distress within the CF population, Angst (2001) reports that more recent research has not demonstrated a difference in the prevalence of emotional problems in CF patients when compared with healthy controls and has in fact pointed to the resilience and adaptability of these patients to their condition. Blair, Cull and Freeman (1994) found that adolescents and young adults with CF were indistinguishable from healthy controls in terms of emotional disturbance and Gee et al (1996) backed this finding with their survey of 90 adults with CF and 106 healthy controls. Gee et al reported that there was no difference in anxiety and depression between the groups although they commented that borderline anxiety was common in both groups as identified on the Hospital Anxiety and Depression Scale. Gee and colleagues also spoke of the

importance of differentiating between specific psychological disorders as opposed to classifying psychological distress as inclusive of both anxiety and depression.

1.3.2 The Influence of Psychological Problems on Adherence

Few studies have investigated the effect of psychological distress on adherence to self-management regimens in chronic conditions, particularly CF. Rapoff (1991) postulates that non-adherence may result from or exist concurrently with emotional problems. Within diabetes literature, research by Kovacs, Goldston, Obrosky and Iyengar (1992) and Jacobson et al (1987) have shown that patients with behavioural or emotional problems are less likely to adhere to diabetic regimens. In the CF population, Gudas et al (1991) reported that greater pessimism and low mood was associated with non-adherence to CF treatment regimens. Furthermore, Koocher, McGrath and Gudas (1990) interviewed 223 patients with CF and their family members and reported that the presence of depression and anxiety interfered with adherence. Both Hains, Davies, Behrens and Biller (1997) and Mullins, Pace and Keller (1994) postulate that cognitive behaviour therapy may be effective to resolve psychological issues, particularly anxiety and depression, in individuals with chronic conditions and consequently improve their adherence to treatment regimens.

1.3.3 Summary

Differences are apparent in reports of the prevalence of psychological distress within the CF population. Whilst some studies claim that there is a high incidence of anxiety and depression in CF patients compared with healthy controls, other studies suggest that there are no differences. Psychological distress has, however, been reported to have a negative impact on adherence to treatment regimens in CF and diabetes

patients. It would therefore seem important that psychological input is available within CF teams in order to resolve these emotional difficulties and reduce their deleterious effect on self-management behaviours.

1.4 SELF-DETERMINATION THEORY OF HUMAN MOTIVATION

1.4.1 Introduction to Self-determination Theory

The Self-determination Theory of Human Motivation, developed by Deci and Ryan (1985), provides a theoretical basis for explaining human motivation to undertake a variety of health related behaviours. The theory maintains that an understanding of motivation to behave requires a consideration of innate psychological needs for competence and autonomy which, when satisfied, enhance self-motivation and well-being and, when thwarted, result in diminished motivation.

1.4.2 Autonomous versus Controlled Professional Relationship Styles

Within a health care setting, self-determination theory emphasises the importance of an 'autonomy supportive' interpersonal climate where a person in a position of authority, for example a physician, takes the perspective of another into account, acknowledges their feelings, solicits their opinions, encourages questions and provides relevant information and opportunities for choice. An autonomy supportive interpersonal style is considered to be closely related to 'patient centredness', a term devised by Laine and Davidoff (1996). Within the health care literature, the terms autonomy and independence have often been used interchangeably but Deci and Ryan (1985) state that autonomous support does not necessarily mean being permissive and giving patients the freedom to make their own medical decisions with little or no advice from professionals. Instead it describes a partnership where the patient is engaged in the discussion, feels their perspective is understood and is provided with information and options about treatment without pressure. In contrast, a controlling style involves pressurising others to behave in particular ways whilst taking little account of their perspective and is considered to be more authoritarian.

The Health Care Climate Questionnaire has been developed by Williams, Freedman and Deci (1998a) to measure patients' perceptions of their health care provider as being autonomy supportive versus controlling whilst discussing self-management behaviours.

1.4.3 Autonomous versus Controlled Motivation

Self-determination theory also distinguishes between different types of motivation. Williams, Saizow and Ryan (1999) report that motivation refers to forces that move people to act. They postulate that some theories of motivation focus exclusively on levels of motivation and state that researchers who think of motivation in this way have tended to examine conditions that result in high levels of motivation versus conditions that result in little or no motivation. However, Williams et al emphasise the importance of differentiating between types of motivation as opposed to simply defining levels of motivation, as they claim that different types of motivation, even when the resulting motivation is high, will lead to very different outcomes. The self-determination theory differentiates between motivation that is 'autonomous' or 'controlled'. Autonomous motivation to behave reflects what people find important and involves a sense of volition and choice over their behaviour whereas controlled motivation depends on explicit or implicit rewards or punishments or people's internalised beliefs about what is expected of them. Controlled motivation also involves the extent to which an individual feels coerced or pressurised to behave or comply in a particular manner. Self-determination theory proposes that behaviour change will occur and persist if it is autonomously motivated in a relevant social context of autonomy supportiveness (Williams, Grow, Freedman, Ryan and Deci, 1996). The Treatment Self-regulation Questionnaire was developed by Ryan and

Connell (1989) to assess the degree to which an individual's motivation for a particular behaviour is autonomous or due to the controlling influence of another.

1.4.4 Self-efficacy

Perceived self-efficacy is a central construct of the Social Cognitive Theory of Human Behaviour (Bandura, 1986). The term describes an individual's belief in their ability to organise and execute the courses of action required in any given task. Rapoff (1999) states that with regard to adherence behaviours to medical regimens, competent functioning requires both skills and self-beliefs of efficacy to use the skills effectively. Rapoff claims that children and adolescents who have the necessary skills and have strong beliefs in their capabilities to perform adherence tasks are more likely to approach difficult regimen tasks as 'challenges' to be mastered rather than 'threats'. Furthermore, they are more likely to set health enhancing goals for themselves and be strongly committed to these goals and increase and sustain their efforts to achieve their goals even when faced with setbacks. Bandura states that perceived self-efficacy is the most powerful determinant of behaviour and O'Leary (1992) postulates that self-efficacy has an influential effect on adherence behaviours, stating that individuals high in self-efficacy are more likely to adhere to medical regimens and thereby improve or maintain their health. Studies undertaken to investigate the influence of self-efficacy on adherence in chronic health conditions have repeatedly demonstrated that self-efficacy is a significant predictor of self-management behaviour.

Within the CF population, Bartholomew, Parcel, Swank and Czyzewski (1993) examined the influence of the self-efficacy of children with CF and their families on

self-management behaviours and concluded that when self-efficacy was enhanced, improvements in the self-management of CF and subsequent health care outcomes was evident. In addition, Parcel et al (1994) reported that self-efficacy was the most important factor in predicting the initiation and continuation of self-management behaviour in CF patients. They stated that interventions that focused on increasing knowledge alone were less effective in improving self-management behaviours in CF patients and their families than interventions that addressed self-efficacy. Williams, Rodin, Ryan, Grolnick and Deci (1998b) posit that self-efficacy is similar to the concept of perceived competence described in the self-determination theory of human motivation and claim that self-efficacy can be enhanced within the context of an autonomy supportive relationship where a health care provider conveys confidence in the individual's ability to achieve negotiated goals. The Self-efficacy Questionnaire was developed by Williams, Freedman and Deci (1998a) to measure an individual's perceived competence in their ability to undertake self-management tasks.

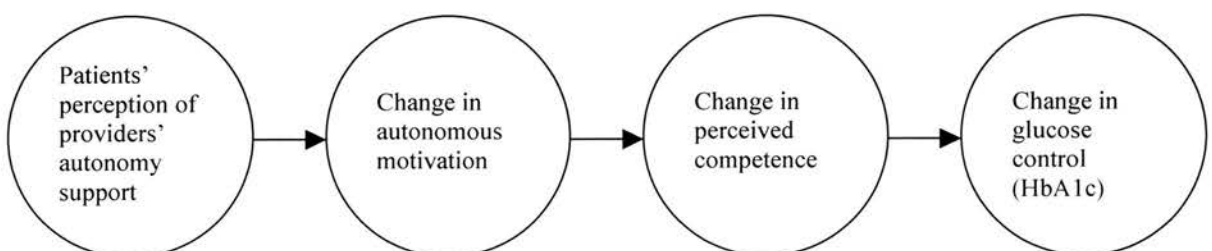
1.4.5 Previous Research Applying Self-determination Theory

Self-determination theory posits that the interpersonal style of significant others will engender different types of motivation. Health care studies guided by the self-determination theory have demonstrated that when health care providers are autonomy supportive, patients are encouraged to develop and maintain more autonomous motivation regarding their health care. Furthermore, patients feel more competent in carrying out a variety of health related behaviours resulting in increased adherence to treatment regimens and improved health outcomes (Deci, Eghrari, Patrick and Leone, 1994). Similar findings have been found in relation to HbA(1c)

reduction for patients with diabetes (Williams et al, 1998a), medication adherence in adult out-patients (Williams et al, 1998b), maintained weight loss in morbidly obese patients (Williams et al, 1996), smoking cessation (Williams and Deci, 1996) and asthma self-management (Howells, Greene, Neville, Pagliari, Mukhopadhyay, Alexander and Greene, 2001).

Williams et al (1998a) investigated whether patients who perceived their health care providers as autonomy supportive would display an increase in autonomous motivation and perceived competence, leading to improved glucose control over a twelve-month study period. 128 individuals between the ages of 18 and 80 years participated in the study and Williams and colleagues found that patients' perception of autonomy support from their health care provider predicted a significant increase in patients' autonomous motivation to regulate their glucose levels. Furthermore, they reported that this increase in autonomous motivation was related to a significant increase in perceived competence in ability to regulate glucose which was in turn related to significant reductions in the HbA(1c) values over twelve months (See Figure 1). Williams et al concluded that attention to psycho-social factors within patient care was as important as consideration to biological contributions to illness since psycho-social factors appeared to significantly influence patients' physical health outcome.

Figure 1: The self-determination model (Williams et al, 1998a)



Williams et al (1998b) also applied the self-determination theory to explore the motivational basis of adherence to long-term medication prescriptions. Participants with various diagnoses who had been taking medication for at least one month completed questionnaires to assess their motivation to adhere and their perceptions of their physicians' interpersonal style. Participants gave subjective ratings of their adherence and a two-day retrospective and a 14-day prospective pill count was undertaken. The results demonstrated that when physicians were perceived by their patients as autonomy supportive, the patients reported greater autonomous motivation for taking their medication and better adherence to prescribed regimens thereby providing further support for the self-determination model. Although Williams et al highlighted the possibility that patient perceptions rather than providers' behaviour were the determining variables, the study was consistent with the hypothesis that autonomy supportiveness on the part of the physician influenced patient behaviour and subsequent health outcomes.

An earlier study by Williams and colleagues (1996) involved 128 morbidly obese patients in a six-month weight loss programme with a 23 month follow up. Patients were asked to rate the health care team as either autonomy supportive or controlling using the Health Care Climate Questionnaire. Williams et al reported that patients who perceived an autonomy supportive interpersonal climate created by the health care staff displayed greater autonomous motivation for weight loss, attended the six-month program more regularly, lost more weight during the program and maintained greater weight loss at follow up than those who perceived the health care team as more controlling. The results of the study demonstrated further support for the self-determination theory. Furthermore, Williams and Deci (1996) conducted a

longitudinal study on smoking cessation and the results suggested that physicians perceived by their patient as autonomy supportive significantly enhanced the autonomous motivation and perceived competence of smokers for not smoking which was related to better cessation over a six-month period. Williams and Deci asserted that the use of an autonomous interpersonal style significantly enhanced smokers' motivation to behave in healthier ways.

In addition, Howells et al (2001) used the self-determination theory to guide their study of 67 young people with asthma, aged between 13 and 22 years. Participants completed the Health Care Climate Questionnaire, Treatment Self-Regulation Questionnaire, Self-efficacy Questionnaire and an Asthma General Knowledge Questionnaire in addition to providing a self-report measure of adherence. The results emphasised the importance of autonomy supportive health care providers and autonomous motivation on improving adherence to asthma self-management with self-efficacy as a potential mediating variable.

1.4.6 Summary

Research on the application of self-determination theory to chronic health conditions is in its early stages, however, results to date have supported the use of the model within health care settings. Consequently, Williams and Deci (2001) posit that physicians should be encouraged to be more autonomy supportive in their interactions with patients since this specific interpersonal style has been associated with more effective health care delivery and more positive health outcomes.

1.5 PRESENT STUDY: AIMS AND HYPOTHESES

1.5.1 Aims

The current study uses the Self-Determination Theory of Human Motivation (Deci and Ryan, 1985) to guide an investigation into factors that promote adherence to CF treatment regimens in adolescents and young adults. The theory highlights the role of autonomy supportive health care provider relationship styles in increasing patients' self-efficacy and motivation for self-management and the present study aims to replicate the findings of previous research in individuals with CF. The study also aims to examine the role of emotional well-being on adherence. As few studies within the CF literature have drawn upon both quantitative and qualitative data, a novel aspect of this research was to obtain detailed information on the personal experiences of young people with CF by using an interview format. In particular, information was gathered regarding the impact of the condition on their lives, reasons for adherence to regimens and particular professional relationship styles associated with good CF self-management. The results of the current study would be expected to provide information regarding factors influential in maximising adherence behaviours that can be presented to health care providers within CF teams to enable them to offer optimal support to young people with CF. The hypotheses are divided into two sections that include predictions from previous research findings and the experimental hypotheses for this study.

1.5.2 Hypotheses

On the basis of previous findings from the literature it is predicted that:

- 1(a) Autonomy supportive relationships between health care providers and adolescents and young adults with CF will be associated with greater autonomous motivation to adhere.
- 1(b) Autonomy supportive relationships between health care providers and adolescents and young adults with CF will be associated with greater self-efficacy.
- 1(c) Autonomous motivation to adhere will be associated with greater self-efficacy.
- 1(d) Adolescents and young adults who report greater self-efficacy will display increased adherence to CF treatment regimens.
- 2 Lower levels of psychological distress (anxiety and depression) will be reported by adolescents and young adults with CF who perceive their health care providers as autonomy supportive.
- 3 Adolescents and young adults with CF who report greater self-efficacy will display lower levels of psychological distress (anxiety and depression).

Experimental hypotheses:

- 4 Higher levels of psychological distress (anxiety and depression) in adolescents and young adults with CF will be associated with poorer adherence to CF treatment regimens.
- 5 Increased age will be associated with better adherence to CF treatment regimens.
- 6 Increased severity of CF will be associated with increased psychological distress (anxiety and depression).

CHAPTER 2: METHOD

2.1 DESIGN

A cross-sectional, investigative study was undertaken with adolescents and young adults attending out-patient CF clinics in Tayside. A pack consisting of five self-report measures was sent to potential participants by post. These measures examined perceived health care provider relationship styles, self-efficacy, motivation to follow a treatment regimen, emotional well-being and adherence behaviour. In addition, semi-structured interviews were carried out with a sub-set of participants to provide qualitative data to augment the quantitative data obtained from the questionnaires.

2.2 PARTICIPANTS

The participants included adolescents and young adults with CF, aged between 12 and 26 years, who attended the out-patient paediatric and adult CF clinics in Tayside. A wide age range was chosen so that any developmental trends could be identified. Within the CF literature there is no consistent classification by which to define adolescents and young adults. In the current study, participants aged between 12-18 years were classified as 'adolescents' whilst participants aged between 19-26 years were classified as 'young adults'. These classifications were applied in line with studies by Angst (2001), Christian and D'Auria (1997) and Duncan-Skingle and Pankhurst (2001) who defined school-aged children as between 7-11 years, adolescents as aged between 12-18 years and young adults as 19 years and upwards respectively. Furthermore, in an attempt to ensure a similar range of ages within each classification, the cut off for young adults was set at 26 years of age.

The criteria for inclusion in the study were:

- Adolescents and young adults with CF aged between 12-26 years
- Current attendance at the CF out-patient clinics in Tayside

The criteria for exclusion in the study were:

- Any individual with CF currently attending psychiatric services
- The presence of another major medical condition

Fifty-three individuals who attended the Tayside CF clinics were invited to participate in the study, 13 adolescents and 40 young adults. Thirty-six individuals did not return the consent forms or questionnaire measures and four individuals contacted the researcher to report that they did not wish to participate in the study as they were undertaking examinations at school. Thirteen individuals agreed to participate in the study. Nine participants completed the questionnaire measures and agreed to be interviewed, although one individual did not attend the appointment for the interview, and four individuals completed the questionnaires only.

Due to the low response rate, the geographical area for recruitment was expanded and additional participants from the Forth Valley CF clinics were invited to participate in the study. An explanation of the steps undertaken in order to do this is presented within the procedure section. Seven individuals, three adolescents and four young adults, were selected and two individuals returned the completed questionnaire pack, bringing the total number of participants to fifteen. Following discussions with the consultants in the Tayside and Forth Valley CF clinics regarding the response rate, the age range of participants was increased to 30 years. Seven

additional individuals met the criteria for inclusion, five in Tayside and two in Forth Valley, bringing the total number of individuals invited to participate in the study to 67. No further responses were returned however.

2.3 MEASURES

The measures employed in the current study are summarised in Table 1 and will be discussed in turn. The questionnaire pack containing these measures can be seen in Appendix 1.

Table 1: Measures used in the current study

<i>VARIABLE MEASURED</i>	<i>MEASURE</i>	<i>REFERENCE</i>
Health care provider relationship style	The Health Care Climate Questionnaire	Williams & Deci (1996)
Self-efficacy	The Self-efficacy Questionnaire	Williams, Freedman & Deci (1998a)
Motivation to adhere to treatment regimens	The Treatment Self-regulation Questionnaire	Ryan & Connell (1989)
Emotional well-being	The Hospital Anxiety and Depression Scale	Zigmond & Snaith (1983)
Adherence	A self-report measure of adherence	Howells, Greene, Neville, Pagliari, Mukhopadhyay, Alexander & Greene (2001)

2.3.1 The Health Care Climate Questionnaire (HCCQ)

The HCCQ was developed by Williams and Deci (1996) to measure patients' perceptions of their health care providers as being 'autonomy supportive' versus 'controlling' in their behaviour towards the patient whilst discussing self-management. The HCCQ consists of fifteen items and responses are measured by a seven-point Likert rating scale ranging from 1 = 'strongly disagree' to 7 = 'strongly agree'. A total score is obtained and the higher the score, the greater the respondent's perception of the health care provider having an 'autonomy supportive' interpersonal style. In the present study the health care providers were represented by the consultant or nurse within the CF clinics. Respondents were notified that they were to answer in terms of their overall perspective of both health care providers.

The HCCQ has been used and validated in studies of weight loss (Williams et al, 1996) and smoking cessation (Williams and Deci, 1996) and internal consistency has been shown to be high with Cronbach α values of 0.92 and 0.96 respectively. Furthermore, Williams and Deci report that patients' perceptions of health care providers are reasonably stable over time with 6-8 month correlations of approximately $r = 0.6$. Although the HCCQ has not been standardised for use with adolescents, Howells et al (2001) modified the layout of the items and response format for use with adolescents with asthma and reported that the items were easily understood and completed by patients. The HCCQ was considered to be appropriate for the current study and permission was obtained from the authors to use the questionnaire. Permission was also obtained from Howells et al to utilise the modified layout and the language was adapted to reflect local CF self-management terminology. A Cronbach α value of 0.83 was obtained in the current study.

2.3.2 The Self-efficacy Questionnaire (SEQ)

This four-item measure was designed by Williams et al (1998a) to assess a person's perceived competence to undertake specific self-management tasks. Responses are scored on a seven-point Likert rating scale ranging from 1 = 'strongly disagree' to 7 = 'strongly agree'. The scale has been used in previous studies involving diabetic and asthmatic patients with good internal consistency as Cronbach α values of 0.80 and 0.94 demonstrate. In the present study the questionnaire was modified with the authors' permission so that it was appropriate for the CF population and a Cronbach α value of 0.88 was obtained.

2.3.3 The Treatment Self-regulation Questionnaire (TSRQ)

The TSRQ was developed by Ryan and Connell (1989) to measure the reasons why people would engage in healthy behaviours, try to change unhealthy behaviours or follow a treatment regimen. The questionnaire assesses the degree to which an individual's motivation for a particular behaviour is autonomous or due to the controlling influence of others, for example, professional carers. The TSRQ is a 13-item measure and respondents rate the degree to which they agree with the items using a seven-point Likert rating scale ranging from 1 = 'strongly disagree' to 7 = 'strongly agree'. Two factors are identified within the measure: Autonomous motivating factors and controlling carers' motivating factors. Questions 2,3,5,7,9,10,11,13 are included within the autonomous motivating factors subscale whilst questions 1,4,6,8,12 make up the controlling carers' motivating factors subscale. An example of an autonomous motivation item is: "I believe that tightly controlling my CF will greatly improve my health" whilst an example of a controlled reason is: "Other people would be mad at me if I didn't".

According to Ryan and Connell the questionnaire has good internal consistency with a Cronbach α value of 0.87 and past research by Williams et al (1996) and Williams et al (1998a) support the construct validity of this measure. In the present study the questionnaire was modified with the authors' permission to include local CF self-management terminology and Cronbach α values of 0.73 and 0.89 were obtained for the autonomous and controlled motivation subscales respectively.

2.3.4 The Hospital Anxiety and Depression Scale (HADS)

The HADS was developed by Zigmond and Snaith in 1983 for use in medical out-patient clinics to detect clinical levels of anxiety and depression, independent of physical symptomatology. The scale consists of 14 items and levels of anxiety and depression are measured on two separate subscales containing seven items each. The respondent is required to rate each item on a four-point rating scale, scored from 0 to 3, so that the total score for each subscale ranges from 0 to 21. The HADS can also be utilised to gauge the severity of anxiety and depression and includes four classifications for score ranges: Normal (0-7), Mild (8-10), Moderate (11-14) and Severe (15-21). A score of 11 or more represents clinical 'caseness' for anxiety or depression.

The reliability and validity of the HADS has been established in studies of medical out-patients (Zigmond and Snaith, 1983) and patients with cancer (Moorey et al, 1991). Zigmond and Snaith report that the HADS demonstrates a high rate of internal consistency with Cronbach α values of 0.93 for the anxiety subscale and 0.9 for the depression subscale. The above studies suggest good psychometric properties based on specific populations such as medical out-patients. The HADS was therefore

considered appropriate to utilise in the current study to detect levels of psychological distress independent of CF symptoms and Cronbach α values of 0.77 and 0.71 were obtained for the anxiety and depression subscales respectively.

2.3.5 Self-report Measure of Adherence

A self-report estimate of adherence was obtained from participants. The measure was developed by Howells et al (2001) for use with asthma patients and was modified for use with CF patients in the current study. Bond and Hussar (1991) assert that patient reports of adherence behaviours are relatively simple and convenient to obtain and Rapoff (1999) states that patient reports address the problem of accessibility to patient behaviours over time and enable estimates to be obtained in ecologically relevant contexts such as the patients' home. Participants were required to estimate their level of adherence to four aspects of CF treatment regimens: Medication (antibiotics), physiotherapy, diet (pancreatic enzymes and vitamin supplements) and exercise. Estimates of adherence were obtained over a one-week period using a five-point Likert rating scale ranging from 0 = 'never/frequently miss days' to 4 = 'every day'. A higher score indicated relative adherence whilst a lower score indicated relative non-adherence. The specified time frame was applied in order to minimise recall errors since Rudd (1993) states that recall for events is generally less than two weeks. Respondents were also asked to comment on whether the previous week had been a 'typical' week to determine whether their estimate of adherence could be considered to be representative of their general adherence to their treatment regimen.

2.3.6 Semi-structured Interview

A semi-structured interview was undertaken to explore the personal experiences of participants on a range of topics that related to the hypotheses of the study. The topics covered during the interview included the impact of CF on the individual's lifestyle, issues relating to adherence or non-adherence to the treatment regimen, emotional well-being and experiences of health care providers' relationship styles. An interview schedule was developed to ensure that all interviews followed a similar outline although the format was flexible to allow the researcher to adapt it depending on the participant's experiences of CF. Participants were also permitted to discuss any issues that had arisen from the completion of the questionnaire measures. Sample questions from the interviews can be seen in Appendix 2. The interview schedule was as follows:

Background Information

- Family background
- Schooling/Employment
- Age at diagnosis

Emotional Well-being

- Feelings about having CF
- Effect of having CF whilst growing up
- Support network

Information Regarding Treatment Regimen

- Treatment regimen required
- Confidence in managing treatment regimen
- Regimen tasks considered the easiest and most difficult to adhere to
- Possible reasons for non-adherence

Relationships with Health Care Providers

- Relationships with health care providers (both positive and negative experiences)
- Satisfaction with the delivery of services

Future Issues

- Worries or concerns about the future/Future health

2.3.7 Other Information Collected

The questionnaire pack issued to participants requested information on age, gender and the health care provider seen most often within the CF clinic. A general information sheet was also developed for the study to record demographic data on age at diagnosis, prescribed treatment regimen, occupation, who the participant lived with and their postcode sector to calculate the corresponding social deprivation category (Carstairs and Morris, 1991). The demographic information was obtained from participants' hospital case notes and the information sheet can be seen in Appendix 3. In addition, an objective measure of physical functioning (FEV1 % predicted) was gathered from the hospital case notes. Lung function, as mentioned earlier, is commonly measured by forced expiratory volume at one second (FEV1) and CF patients are assigned a predicted FEV1 value based on their gender, age and height. The FEV1 % predicted gives an indication of the capacity to which an individual is functioning, for example at 70% of their predicted FEV1. Since lung function can deteriorate with infections, a higher FEV1 indicates a stable period of health (Ramsey and Marshall, 1995). FEV1 % predicted is commonly used in clinical and research settings as a measure of severity of illness and in the current study an average FEV1 % predicted value was calculated for each participant from readings obtained during the previous six months.

2.4 PROCEDURE

Participants were initially recruited from patients currently attending the paediatric and adult CF out-patient clinics in a large teaching hospital in Tayside. The researcher met with the consultant paediatricians, the consultant respiratory physicians and the CF liaison nurses to discuss the nature of the study and gain their approval to proceed. Ethical approval was then sought and obtained from the Tayside Committee on Medical Research Ethics. No changes to the research design were required but minor changes were requested for the participants' introductory letter and information sheet. A detailed account of the time delay incurred whilst awaiting ethical approval from the research ethics committee is presented within the discussion section. Caldicott Guardian approval to access participants' hospital case notes was also obtained from the Medical Directors within Tayside NHS Health Board and thereafter, suitable participants were identified by the consultants. To maintain confidentiality the researcher was not permitted to access patient details until the individuals had consented to participate in the study. The CF liaison nurses therefore agreed to send out by post the introductory letter, information sheet detailing the study, consent form and questionnaire pack to the selected individuals (See Appendices 4,5,6 and 1 respectively). A reply slip was also enclosed for completion by individuals who were willing to attend the semi-structured interview (See Appendix 7).

Individuals were encouraged to contact the researcher if they had any questions about the study or required further information prior to giving informed consent. They were asked to decide whether or not to participate in the study within a fortnight and to return the completed questionnaire pack in the self-addressed

envelope provided. Individuals were informed that their decision to participate in the research was entirely voluntary and would not affect their treatment if they chose to withdraw from the study at any time. Confidentiality of responses was assured although the information sheet specified circumstances when information would be fed-back to the consultants with the participant's consent, for example, if any participant appeared to be exhibiting significant emotional distress from their answers to the HADS with a score of 11+ on either subscale. Individuals were also notified that their GP would be informed of their participation in the study (See Appendix 8 for letter sent to GPs). Additional parental consent was sought for those individuals under 16 years of age and an introductory letter and information sheet for parents was enclosed with the correspondence (See Appendices 9 and 10). Three weeks after the initial correspondence was issued, a reminder letter was sent to all participants or their parent/guardian to encourage the return of further consent forms and completed questionnaire packs and this letter is included in Appendix 11.

Due to the low response rate, the researcher contacted the consultant respiratory physicians in two hospitals in Forth Valley to discuss the study. Thereafter, a proposal was submitted to the Forth Valley Ethics of Research Committee and following minor changes, approval was granted for the study to be undertaken. Caldicott Guardian approval to access participants' hospital case notes was also obtained from the Medical Director of Forth Valley NHS Health Board. The CF liaison nurses agreed to send out the introductory letter, information sheet, consent form and questionnaire pack, shown in Appendices 4,5,6 and 1 respectively, to those individuals considered suitable for inclusion by the consultants. A reminder letter was also sent out three weeks after the initial correspondence to elicit further

responses (See Appendix 11). Following additional discussions with the consultants within the Tayside and Forth Valley CF clinics regarding the response rate, it was decided that the age range of participants would be increased to 30 years and the Tayside Committee on Medical Research Ethics and Forth Valley Ethics of Research Committee were notified of and approved this amendment.

Having received the completed questionnaire measures the researcher telephoned those participants who had agreed to be interviewed in order to arrange a convenient appointment. All interviews were conducted by the researcher within the participants' homes and lasted between 40 minutes to an hour. Where possible, the interviews were undertaken within two weeks of receiving the completed questionnaire packs from each participant. All interviews were tape recorded with participants' consent and were fully transcribed afterwards. So as not to influence the course of the interview, the researcher reviewed participants' hospital case notes to collect the required demographic details after completing the interview.

2.5 ANALYSIS OF DATA

2.5.1 Data Analysis

The data was analysed using the Statistical Package for Social Sciences (SPSS) for Windows 2000, PC-version 10.0. All variables were checked for assumptions of normality by examining frequency distribution charts and appropriate statistical tests were selected accordingly. Descriptive statistics and an Independent t-test were employed on the demographic data and relationships between variables were calculated using parametric bivariate correlations (Pearson r correlation co-efficient) and partial correlations. A larger sample size would have allowed for multiple

regression analysis to examine potential predictors of adherence to CF self-management.

A thematic analysis of the interview data was undertaken to explore the impact of CF on the participants' lifestyle, issues affecting adherence to treatment regimens and experiences of health care providers' interpersonal styles. Repeated readings of the transcripts were undertaken to identify themes and patterns of responses. Current literature examining adherence to treatment regimens in chronic health conditions, the influence of health care professional relationship styles on adherence and emotional well-being in chronic conditions was also consulted to facilitate the development of themes for the analysis.

2.5.2 Statistical Power

Following discussions with the consultant paediatricians and consultant respiratory physicians in Tayside it was expected that approximately 60 patients would meet the inclusion criteria for the study. In view of results from a similar study by Howells et al (2001) within the asthma population, a large effect size was anticipated. A power calculation indicated that at least 40 participants would be required in order to achieve sufficient statistical power to detect a large effect size at the 0.8 level (Cohen, 1992). Fewer participants were recruited than was initially anticipated, however, and possible reasons for the low response rate are identified within the discussion section. Post-hoc power calculations can also be seen within the discussion section.

CHAPTER 3: RESULTS

3.1 DEMOGRAPHIC DATA

3.1.1 Exploration of the Data

The data was explored prior to statistical analysis to check for normality and those variables showing significant skewness or kurtosis were transformed. The only variable found to depart from normality was age at diagnosis which was positively skewed. A logarithm (to the base 10) transformation was therefore applied so that the values of the variable more closely matched the normal distribution thereby satisfying the distribution requirements for the use of parametric statistics. Throughout this section, significant results are reported at the $p < 0.05$ level, or $p < 0.01$ where applicable, and all significant results are reported in bold type.

3.1.2 Participants

Sixty-seven individuals were invited to participate in the study. Fifty-eight individuals were selected from the Tayside CF clinics with nine individuals selected from the Forth Valley CF clinics. Forty-eight individuals (71.6%) did not return the consent forms or questionnaire measures and four individuals (6%) contacted the researcher to report that they did not wish to participate in the study as they were undertaking examinations at school. Fifteen individuals, seven males and eight females, were included in the study, giving a response rate of 22.4%. Nine participants completed the questionnaire measures and agreed to be interviewed, although one individual did not attend the appointment for the interview, and six individuals consented to complete the questionnaires only. A summary of the number of patients approached and recruited from the CF clinics within Tayside and Forth Valley Health Board is shown in Table 2.

Table 2: Summary of patients approached and recruited (n=67)

	TAYSIDE	FORTH VALLEY
Eligible participants approached	58	9
Completed questionnaires only	5 (8.6%)	2 (22.2%)
Completed both questionnaires and interview	8 (13.8%)	0
Declined to participate	4 (6.9%)	0
Did not respond	41 (70.7%)	7 (77.8%)

3.1.3 Summary of Demographic Data

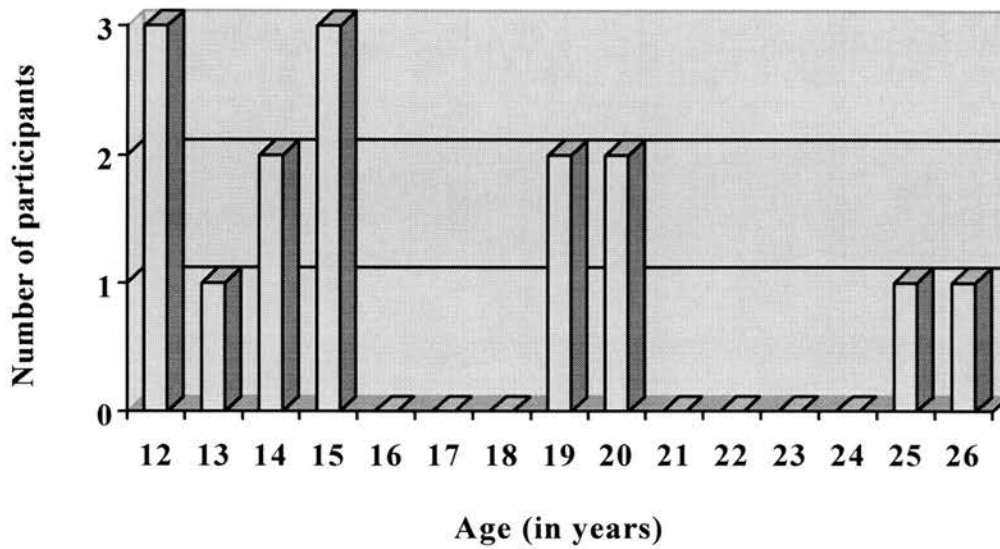
On examination the two respondents from the Forth Valley CF clinics did not appear to differ from the Tayside sample on any of the demographic variables (See Appendix 12 for graphs of participants' demographic details). It was therefore considered appropriate to combine all participants' responses during the data analysis. A summary of demographic details of participants from the CF clinics within Tayside and Forth Valley Health Boards is given in Table 3. The table includes gender, age (in years), age at diagnosis (in months), severity of illness and social deprivation category (Depcat). Participants' postcode sectors were used to calculate the corresponding social deprivation category ranging from Depcat 1 and 2 = 'relatively affluent area' to Depcat 6 and 7 = 'highly deprived area' (Carstairs and Morris, 1991).

Table 3: Demographic details of participants (n=15)

	Participants from Tayside and Forth Valley				
Variable	N	Mean	SD	Min.	Max.
Female	8				
Male	7				
Age (in years)		16.73	4.56	12	26
Age at diagnosis (in months)		16.13	30.61	1	120
Severity of illness (FEV1% predicted)		77.33	13.96	52	99
Social deprivation category (Depcat 1-7)		3.60	1.35	2	6

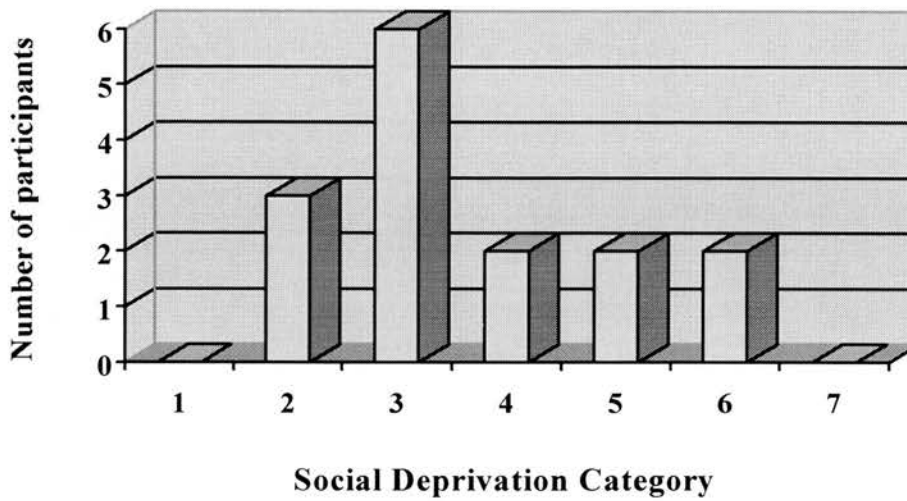
Non-respondents: In order to maintain confidentiality, the Medical Directors within Tayside and Forth Valley NHS Health Boards granted Caldicott Guardian approval to access participants' hospital case notes on the understanding that the case notes would not be accessed unless consent to participate in the study had been received. It was therefore not possible to gather any demographic information about non-responders in order to assess whether the responses obtained were representative of the CF population as a whole.

Age: The frequency distribution of ages of participants is shown in Figure 2. Nine participants (60%) were classified as adolescents (12-18 years) and six participants (40%) were classified as young adults (19-26 years).

Figure 2: Age of participants (n=15)

Age at diagnosis: With regard to age at diagnosis, eleven participants (73.3%) were diagnosed with CF within the first six months following birth. Two participants (13.3%) were diagnosed by two years of age, one participant (6.7%) was diagnosed at three years of age and the remaining participant (6.7%) was not diagnosed until ten years of age.

Social Deprivation Category: The frequency distribution of social deprivation categories for all participants is illustrated in Figure 3. Three participants (20%) came from Depcat 2. The majority of participants came from Depcat 3 and 4 with six participants (40%) from Depcat 3 and two participants (13.3%) from Depcat 4. Two participants (13.3%) came from Depcat 5 and two participants (13.3%) came from Depcat 6. No participants lived in the most or least affluent areas (Depcat 1 and 7 respectively).

Figure 3: Social deprivation categories for all participants (n=15)

Living situation: With regard to the living situation of participants, twelve participants (80%) lived with their parents, two participants (13.3%) lived with their partner and one participant (6.7%) lived alone.

Working status: In relation to the working status of participants, nine participants (60%) were attending school, three participants (20%) were in higher education, one participant (6.7%) was in part-time work and two participants were not working due to ill health (13.3%).

Treatment regimen: All participants reported that they were required to undertake the four specified CF treatment regimen tasks including medication, physiotherapy, diet and exercise on a daily basis. With regard to the self-report estimate of adherence over a one-week period, all participants reported that the previous week had been a 'typical' week with no major life events reported. It was therefore considered that their estimate of adherence over the one-week period was representative of their general adherence to the treatment regimen.

3.1.4 Summary of Questionnaire Data

A summary of data obtained from the five questionnaire measures completed by participants is shown in Table 4. The table includes the means, standard deviations and range of scores on the measures.

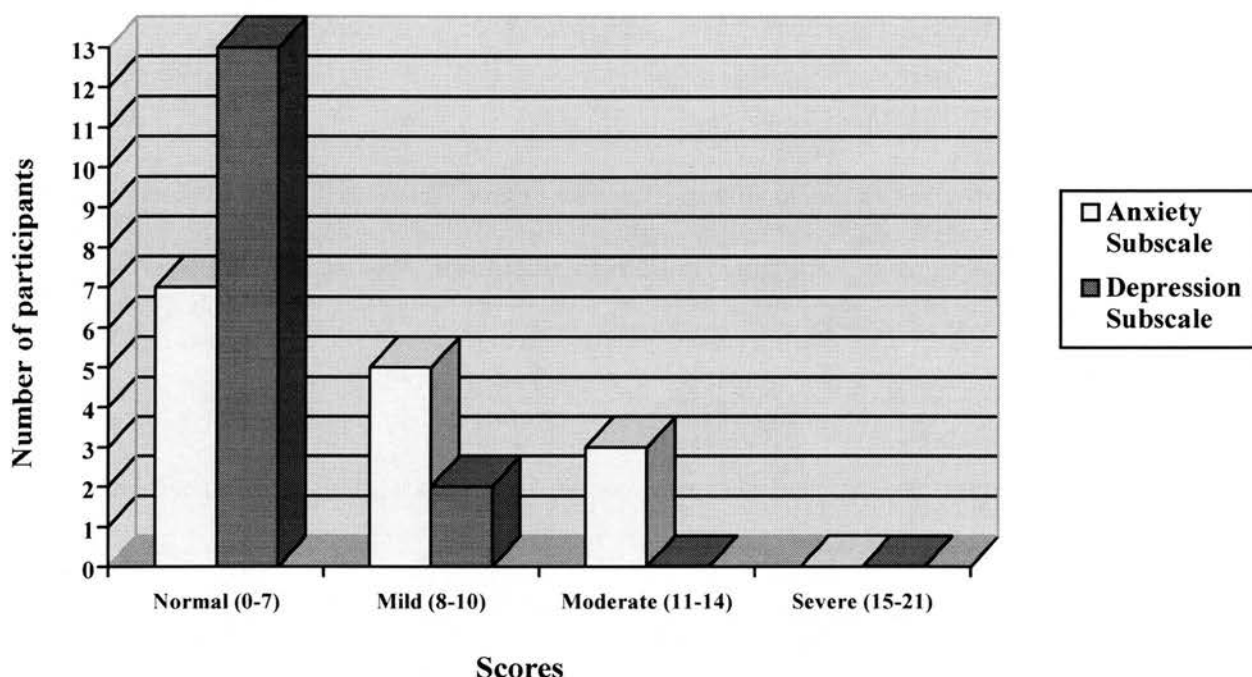
Table 4: Details of responses to questionnaire measures

Variables	N	Mean	SD	Min.	Max.
The Health Care Climate Questionnaire	15	81.73	11.56	62	99
The Self-efficacy Questionnaire	15	24.27	3.77	18	28
The Treatment Self-regulation Questionnaire					
- Autonomous Subscale	15	45.13	6.99	33	55
- Controlled Subscale	15	23.60	8.56	9	35
The Hospital Anxiety and Depression Scale					
- Anxiety Subscale	15	6.93	4.10	0	13
- Depression Subscale	15	3.47	3.11	0	9
The Self-report Adherence Measure					
- Adherence to Medication	15	3.30	.528	2	4
- Adherence to Physiotherapy	15	2.10	1.18	.5	4
- Adherence to Diet	15	3.40	.687	2	4
- Adherence to Exercise	15	3.00	.964	1	4

Emotional Well-being: A frequency distribution of scores for all participants on the Hospital Anxiety and Depression Scale is illustrated in Figure 4. On the anxiety subscale, seven participants (46.7%) fell within the ‘normal’ range of scores (0-7), five participants (33.3%) fell within the ‘mild’ range of scores (8-10) and three participants (20%) fell within the ‘moderate’ range of scores (11-14), demonstrating clinical ‘caseness’ for anxiety. No participants presented with ‘severe’ levels of

anxiety (15-21). On the depression subscale, thirteen participants (86.7%) fell within the ‘normal’ range of scores (0-7) and two participants (13.3%) fell within the ‘mild’ range of scores (8-10). No participants presented with ‘moderate’ or ‘severe’ levels of depression.

Figure 4: Scores on the anxiety and depression subscales for all participants (n=15)



Relationship between gender, social deprivation category, severity of illness and adherence to cystic fibrosis treatment regimen tasks: An Independent t-test was conducted to investigate whether participants’ gender had any effect on their adherence. In addition, Pearson’s Correlations were undertaken to examine whether there were any relationships between social deprivation category, severity of illness and adherence to self-care components. The results are displayed in Tables 5, 6 and 7.

Table 5: Independent t-test examining effect of gender on participants' adherence to cystic fibrosis treatment regimen tasks

	Mean scores		t	df	Sig. (one-tailed)
	Male	Female			
Adherence to Medication	3.36	3.25	.380 (p=.355)	13	(NS)
Adherence to Physiotherapy	1.93	2.25	-.511 (p=.309)	13	(NS)
Adherence to Diet	3.36	3.44	-.218 (p=.415)	13	(NS)
Adherence to Exercise	2.86	3.13	-.523 (p=.305)	13	(NS)

Results indicate that there is no significant difference between male and female participants on their scores on the measure of adherence to the four cystic fibrosis treatment regimen tasks.

Table 6: Correlation comparing social deprivation category with participants' adherence to cystic fibrosis treatment regimen tasks

	Social Deprivation Category		
	N	Pearson's Correlation	Sig. (one-tailed)
Adherence to Medication	15	-.270 (p=.165)	(NS)
Adherence to Physiotherapy	15	-.018 (p=.475)	(NS)
Adherence to Diet	15	-.431 (p=.054)	(NS)
Adherence to Exercise	15	-.384 (p=.079)	(NS)

There was no significant relationship between social deprivation category and adherence to cystic fibrosis treatment regimen tasks.

Table 7: Correlation comparing severity of illness with participants' adherence to cystic fibrosis treatment regimen tasks

	Severity of Illness		
	N	Pearson's Correlation	Sig. (one-tailed)
Adherence to Medication	15	-.097 (p=.366)	(NS)
Adherence to Physiotherapy	15	.285 (p=.151)	(NS)
Adherence to Diet	15	-.257 (p=.177)	(NS)
Adherence to Exercise	15	.122 (p=.332)	(NS)

There was no significant relationship between severity of illness and adherence to cystic fibrosis treatment regimen tasks.

3.2 HYPOTHESES FROM THE LITERATURE

3.2.1 Hypothesis 1(a)

Autonomy supportive relationships between health care providers and adolescents and young adults with CF will be associated with greater autonomous motivation to adhere. Research has suggested that patients display greater autonomous motivation to adhere when they perceive their health care providers as being autonomy supportive. A Pearson’s Correlation was conducted to test for a relationship between the scores on the measures of health care providers’ relationship style and participants’ motivation to adhere as shown in Table 8.

Table 8: Correlation comparing health care providers’ relationship style with participants’ motivation to adhere

	Health Care Providers’ Relationship Style		
	N	Pearson’s Correlation	Sig. (one-tailed)
Motivation to Adhere	15	.566 (p=.014)	p<0.05

Results indicate that there is a meaningful relationship between autonomy supportive health care provider relationship styles and participants’ autonomous motivation to adhere ($r = .566$; $n=15$; $p<0.05$). An additional correlation was conducted between scores on the measures of health care providers’ relationship style and participants’ controlled motivation to adhere. As anticipated, no significant correlation was found between autonomy supportive health care provider relationship styles and participants’ controlled motivation ($r = .206$; $n=15$; $p = .231$).

3.2.2 Hypothesis 1(b)

Autonomy supportive relationships between health care providers and adolescents and young adults with CF will be associated with greater self-efficacy. Researchers have suggested that participants are more likely to display increased self-efficacy when they perceive their health care provider as autonomy supportive. Scores on the measure of perceived health care providers' relationship style were compared with scores on the measure of self-efficacy. A Pearson's Correlation was conducted to test for a relationship between these variables as illustrated in Tables 9.

Table 9: Correlation comparing health care providers' relationship style with participants' self-efficacy

	Health Care Providers' Relationship Style		
	N	Pearson's Correlation	Sig. (one-tailed)
Self-efficacy	15	.689 (p=.002)	p<0.01

Results indicate a meaningful correlation between autonomy supportive health care provider relationship styles and participants' self-efficacy ($r = .689$; $n=15$; $p<0.01$).

3.2.3 Hypothesis 1(c)

Autonomous motivation to adhere will be associated with greater self-efficacy. Research suggests that when participants display autonomous motivation to adhere, they will also display greater levels of self-efficacy. Scores of participants' motivation to adhere were compared with scores on the measure of self-efficacy. This analysis was conducted using a Pearson's Correlation as shown in Table 10.

Table 10: Correlation comparing participants’ motivation to adhere and self-efficacy

	Motivation to Adhere		
	N	Pearson’s Correlation	Sig. (one-tailed)
Self-efficacy	15	.478 (p=.036)	p<0.05

Results indicate that there is a meaningful relationship between participants’ autonomous motivation to adhere and their self-efficacy ($r = .478$; $n=15$; $p<0.05$). An additional correlation was conducted between scores on the measure of self-efficacy and participants’ controlled motivation to adhere. As anticipated, no significant correlation was found between self-efficacy and participants’ controlled motivation ($r = .294$; $n=15$; $p = .144$).

3.2.4 Hypothesis 1(d)

Adolescents and young adults who report greater self-efficacy will display increased adherence to CF treatment regimens. Research has indicated that greater self-efficacy results in improved adherence. Scores on the measure of self-efficacy were compared with scores of adherence to each of the four individual CF treatment regimen tasks. A Pearson’s Correlation was carried out to compare scores as illustrated in Table 11.

Table 11: Correlation comparing participants' self-efficacy with adherence to cystic fibrosis treatment regimen tasks

	Self-efficacy		
	N	Pearson's Correlation	Sig. (one-tailed)
Adherence to Medication	15	.316 (p=.126)	(NS)
Adherence to Physiotherapy	15	.282 (p=.154)	(NS)
Adherence to Diet	15	.466 (p=.040)	p<0.05
Adherence to Exercise	15	.098 (p=.364)	(NS)

Results indicate that there is a meaningful relationship between participants' self-efficacy and adherence to diet in adolescents and adults with CF ($r = .466$; $n=15$; $p<0.05$). No significant correlations were found between participants' self-efficacy and their adherence to medication, physiotherapy or exercise.

The results provide some evidence that the self-determination model of human motivation could be applicable within the CF population, since meaningful relationships were found between autonomy supportive health care providers and participants' autonomous motivation to adhere and between participants' autonomous motivation to adhere and participants' self-efficacy. Furthermore, there was a meaningful correlation between participants' self-efficacy and their adherence to diet. As there was a meaningful correlation between autonomy support from health care providers and participants' self-efficacy, a partial correlation was conducted controlling for participants' autonomous motivation to adhere to establish whether

autonomous motivation was a mediating variable between autonomy support and self-efficacy. The results of the partial correlation are presented in Table 12.

Table 12: Partial correlation comparing health care providers’ relationship style with participants’ self-efficacy whilst controlling for participants’ autonomous motivation to adhere

	Health Care Providers’ Relationship Style		
	Partial Correlation	df	Sig. (one-tailed)
Self-efficacy	.577 (p=.015)	12	p<0.05

The correlation between health care providers’ relationship style and participants’ self-efficacy, when controlling for participants’ autonomous motivation, was less significant ($r = .577$; $df = 12$; $p<0.05$). The results therefore suggest that participants’ autonomous motivation to adhere is a mediator in the link between autonomy supportive health care providers and participants’ increased self-efficacy.

3.2.5 Hypothesis 2

Lower levels of psychological distress (anxiety and depression) will be reported by adolescents and young adults with CF who perceive their health care providers as autonomy supportive. Research indicates that individuals who perceive their health care provider as being autonomy supportive will display better psychological adjustment. Scores on the measure of perceived health care providers’ relationship style were compared with scores on the measure of psychological well-being. This analysis was carried out using a Pearson’s Correlation as shown in Table 13.

Table 13: Correlation comparing health care providers’ relationship style with participants’ psychological well-being

Psychological Well-being	Health Care Provider’s Relationship Style		
	N	Pearson’s Correlation	Sig. (one-tailed)
Anxiety	15	-.323 (p=.120)	(NS)
Depression	15	-.298 (p=.140)	(NS)

No significant correlations were found between autonomy supportive health care providers and participants’ levels of anxiety and depression, although the results suggest a trend in the hypothesised direction with lower levels of anxiety and depression being experienced if health care providers are perceived as autonomy supportive.

3.2.6 Hypothesis 3

Adolescents and young adults with CF who report greater self-efficacy will display lower levels of psychological distress (anxiety and depression). Research indicates that individuals with high self-efficacy display better psychological adjustment. Scores on the measure of self-efficacy were compared with scores on the measure of psychological well-being. This analysis was carried out using a Pearson’s Correlation as shown in Table 14.

Table 14: Correlation comparing participants’ self-efficacy with their psychological well-being

	Self-efficacy		
Psychological Well-being	N	Pearson’s Correlation	Sig. (one-tailed)
Anxiety	15	-.438 (p=.051)	(NS)
Depression	15	-.237 (p=.198)	(NS)

No significant correlations were found between participants’ self-efficacy and their levels of anxiety and depression, although the results suggest a trend in the hypothesised direction with lower levels of anxiety, in particular, being experienced if participants’ self-efficacy is high.

3.3 EXPERIMENTAL HYPOTHESES

3.3.1 Hypothesis 4

Higher levels of psychological distress (anxiety and depression) in adolescents and young adults with CF will be associated with poorer adherence to CF treatment regimens. Researchers indicate that emotional distress can have a negative impact on adherence to treatment regimens. Scores on the measure of psychological well-being were compared with scores of adherence to each of the four individual CF treatment regimen tasks. Pearson’s Correlations were conducted as shown in Table 15 and 16.

Table 15: Correlation comparing levels of anxiety with adherence to cystic fibrosis treatment regimen tasks

	Anxiety		
	N	Pearson's Correlation	Sig. (one-tailed)
Adherence to medication	15	-.122 (p=.332)	(NS)
Adherence to physiotherapy	15	.090 (p=.375)	(NS)
Adherence to diet	15	-.168 (p=.275)	(NS)
Adherence to exercise	15	.208 (p=.228)	(NS)

Table 16: Correlation comparing levels of depression with adherence to cystic fibrosis treatment regimen tasks

	Depression		
	N	Pearson's Correlation	Sig. (one-tailed)
Adherence to medication	15	-.200 (p=.237)	(NS)
Adherence to physiotherapy	15	.277 (p=.159)	(NS)
Adherence to diet	15	-.361 (p=.093)	(NS)
Adherence to exercise	15	-.095 (p=.368)	(NS)

The results suggest no significant correlations between participant's levels of anxiety and depression and their adherence to the individual CF treatment regimen tasks.

However, the results suggest a trend in the anticipated direction particularly for adherence to diet, with poorer adherence to this individual regimen task occurring if higher levels of depression are experienced ($r = -.361; n=15; p = .093$).

3.3.2 Hypothesis 5

Increased age will be associated with better adherence to CF treatment regimens. Research in chronic health conditions has suggested that adolescence is linked with poorer adherence to treatment regimens. It was therefore hypothesised that the younger the age of the sample, the poorer the adherence. Age was compared with scores of adherence on each of the CF treatment regimen tasks. This analysis was conducted using a Pearson's Correlation as illustrated in Table 17.

Table 17: Correlation comparing age with adherence to cystic fibrosis treatment regimen tasks

	Age		
	N	Pearson's Correlation	Sig. (one-tailed)
Adherence to medication	15	-.024 ($p=.467$)	(NS)
Adherence to physiotherapy	15	-.491 ($p=.031$)	$p<0.05$
Adherence to diet	15	.367 ($p=.089$)	(NS)
Adherence to exercise	15	.228 ($p=.207$)	(NS)

Results suggest that there is a meaningful negative correlation between adherence for physiotherapy and age ($r = -.491$; $n=15$; $p<0.05$). No significant correlations were found between age and adherence to medication, diet or exercise.

3.3.3 Hypothesis 6

Increased severity of CF will be associated with increased psychological distress (anxiety and depression). It was hypothesised that adolescents and young adults with CF will display increased levels of psychological distress as their condition deteriorates. Scores on an objective measure of severity of illness (FEV1 % predicted) were compared with scores from the measure of psychological well-being. A Pearson’s Correlation was carried out to compare the scores as shown in Table 18.

Table 18: Correlation comparing severity of illness with participants’ Psychological Well-being

	Severity of Illness		
	N	Pearson’s Correlation	Sig. (one-tailed)
Anxiety	15	-.336 (p=.111)	(NS)
Depression	15	-.135 (p=.315)	(NS)

No significant correlation was found between severity of condition and levels of anxiety or depression in adolescents and young adults with CF. However, results indicate a trend in the anticipated direction for anxiety with increased anxiety being experienced with a deterioration in the severity of illness ($r = -.336$; $n=15$; $p = .111$).

3.4 QUALITATIVE DATA

3.4.1 Participants

All sixty-seven individuals approached to participate in the main study were invited to attend a semi-structured interview with the researcher following the completion of the questionnaire measures. Of the fifteen individuals who returned the questionnaire measures, nine individuals agreed to attend an interview, although one individual failed to attend the arranged appointment. Consequently eight participants were interviewed for the study. All the interviews were tape-recorded with participants' consent and were fully transcribed afterwards. A summary of the participants who attended the interview is given below in Table 19.

Table 19: Descriptive information on participants who attended the semi-structured interviews (n = 8).

Respondent	Gender	Age (In years)	Age at Diagnosis (In months)	Severity of CF (FEV1 % predicted)	Current Treatment (Medication, physiotherapy, diet, exercise)
1	Male	12	36	65	All the above
2	Female	14	1	85	All the above
3	Male	12	2	80	All the above
4	Female	14	3	99	All the above
5	Female	19	5	83	All the above
6	Female	19	2	83	All the above
7	Male	26	24	53	All the above
8	Female	15	1	90	All the above

3.4.2 Researcher Bias

It is important to outline researcher bias before the analysis of the interview data. In the current study, researcher bias was directed towards factors influencing adherence to treatment regimens and reasons for non-adherence. In addition, the bias that health care providers perceived by participants as autonomy supportive would influence participants' motivation to adhere and self-efficacy was also evident.

3.4.3 Data Analysis

A thematic analysis of the interview data was undertaken for each respondent. Repeated readings of interview transcripts were carried out to identify themes and similar experiences and events were grouped together in categories. Current literature on adherence in CF, psychological well-being in chronic illnesses and self-determination theory was also consulted to facilitate the development of themes for the analysis. The main themes explored were taken from research by Rapoff (1999), Angst (2001) and Williams et al (1998a). Due to time constraints a more thorough analysis of the data using a grounded theory approach was not conducted but the most relevant findings are presented.

3.4.4 Themes Identified

The most salient themes identified that corresponded to the aims of the study were split into four broad categories as outlined in Figure 5.

Figure 5: The main categories identified from the interview transcripts that correspond to the aims of the study

Impact of Cystic Fibrosis on Participants' Lifestyles
Effect of frequent hospital admissions
Effect of CF on schooling
Effect of CF on employment
Relationships and fertility issues
Concerns about future health
Emotional Well-being in Participants
Emotional reactions to CF (Anxiety, low mood, embarrassment and frustration)
Importance of support of significant others (Emotional and practical support)
Importance of acceptance of the condition and maintaining a sense of normality
Adherence Issues
Reasons for adherence
Reasons for non-adherence
Most difficult treatments to adhere to
Relationships with Health Care Providers
Satisfaction with the CF team
Negative experiences with health care providers
Importance of autonomy in treatment
Confidence in undertaking treatments

Examples from each of the above categories are presented overleaf. For each theme, respondents who mentioned the particular theme are identified in brackets.

3.4.4.1 Impact of Cystic Fibrosis on Participants' Lifestyles

Effect of Frequent Hospital Admissions

Five respondents discussed the impact of frequent hospitalisations on their life. Three of the individuals reported missing their family whilst in hospital and one respondent in particular reported feeling isolated as they required to be segregated from the other CF patients within the ward because they had *Burkholderia cepacia*. Two respondents reported that they enjoyed being in hospital as it was an opportunity to meet up with their peers with CF.

"I came out of hospital about three weeks ago...was getting IV's. I don't like going into hospital, it's boring, there is nothing to do...I miss my mum and family but mum visits every day and its great to get home and see everyone"

"I don't like being in hospital too often...I'm not allowed in the bay with the other CF children as I have *Burkholderia cepacia*...I miss my family" (2,3,8)

"My friend is usually in hospital when I am...it's more fun...we share a room" (1,7)

Effect of CF on Schooling

The majority of respondents reported having missed a substantial amount of school due to their condition. However, it appeared that the schools had been accommodating by permitting the individuals to come in later in the mornings, so that they could complete their morning treatment regimens, and sending work home during long periods of ill health in an attempt to prevent the respondents from falling behind with their academic work. Two respondents commented on parental concerns that they may be bullied about their CF within the school environment although they indicated that they had encountered no such problems.

“I was off sick about half the time I was at school...school sent me homework if I was off for long periods. Even if I was in hospital there was a teacher there so I didn't get out of working even if I was ill”

“I'm on a half-time curriculum because of my CF...if I'm off for a while I get sent homework”

“I'm off a lot...it's not good because I miss the work” (1,2,3,5,7,8)

“School make allowances for me getting time off school for appointments or coming in later in the mornings because of my treatments” (4,5,6)

“Mum and dad were worried that I would be bullied about my CF but I never was”

“I never felt segregated or embarrassed or anything like that at school...mum and dad worried I may be bullied or something because of the CF but I wasn't” (2,6)

Effect of CF on Employment

The three young adults who attended the interview commented on the effect of CF on their choice of employment. One adolescent discussed their parents' lowered expectations of their future employment.

“Originally I wanted to be a mid-wife or nurse but I wasn't allowed to do that because of the risk of cross infection. I'm doing primary school teaching now at college...it's a sense of achievement”

“I went to college to do a catering course and got a job in an industrial kitchen but it was too hot and when I'd leave at night and go outside the air was cold and I got a lot of chest infections. I had to give it up as I couldn't work in the kitchens...so I wasted the two years at college” (5,6,7)

“Mum is not as ambitious for me career wise as she is for my sisters. All she wants is for me to remain healthy” (4)

Relationships and Fertility Issues

All of the young adults reported that they were currently in a relationship with a supportive partner. One of the young adults discussed concerns regarding their ability to cope with pregnancy in view of their condition and another spoke of the impact of being infertile on a previous relationship.

“I have been living with my boyfriend for six years, he’s very supportive about my CF” (5,6,7)

“I do have some worries about starting a family. How will I cope with the pregnancy? I also worry about my child’s health and the effect of me having CF on my children” (6)

“I was engaged once but my fiancé got pregnant. There were questions about my fertility because of the CF...I found out I was not the father...the relationship ended” (7)

Concerns about Future Health

Five respondents reported that they did not think about their future health. Three of these individuals reported that they had no concerns since their CF was relatively well controlled at present, compared to some of their peers with CF. The other two individuals commented on the possible impact that their peer-related activities may have on their health.

“I don’t think about it, I just get on with my life”

“I don’t worry about what will happen as I get older, I don’t think about it...I live each day as it comes” (2,4,5,6,7)

“I don’t have any worries about my health in the future...I have kept healthy to date”

“I’m more healthy than lots of other people with CF I know” (2,5,6)

“My liver is not in good shape. I’ll have to watch with alcohol. Mum keeps saying I can’t binge at parties or I’ll damage my liver”

“My asthma has been bothering me at the moment...I need to use the inhaler a lot which gets annoying. It’s probably because I smoke a lot. I keep getting told to stop and give up...I managed for a few months but it’s really difficult because if I’m at the pub with my pals and they are smoking it’s really hard not to join them” (4, 7)

One of the respondents who reported having no current concerns about their future health stated that this had not always been the case.

“I had an older sister but she died of CF 10 years ago this year...she was very ill. I found it hard at times after her death...I was worried about having CF and worried about my health. I concentrated on my physio and medication for a few years afterwards so that my condition did not worsen” (6)

3.4.4.2 Emotional Well-being in Participants

Emotional Reactions to CF

A variety of emotions relating to the condition were reported by all respondents. Two respondents commented that maintaining a positive mood played an important part in helping them manage their condition.

“My nature helps me to cope with CF...its my nature to stay positive”

“I try to stay positive and not let it (CF) affect my life” (2,4)

Five respondents reported experiencing problems with anxiety or low mood as a result of their condition. Concerns about the implications of poor weight gain and the need for regular intravenous antibiotics were given as particular examples.

“I’m not gaining weight or height...it’s worrying. The doctors suggested that I needed a peg tube so that I can get overnight feeds but I don’t want one inserted as I’ve had enough operations... I don’t want a tube in my stomach” (1)

“When I get IV’s I do get fed up at times and feel down. You can’t stay with friends or have a shower or dress easily. IV’s are more invasive...you depend on your family and friends at these times to pick you back up when you feel low” (2,4,6,7)

Five respondents described feelings of embarrassment in social situations because of their CF-related symptoms and treatments, particularly the need to take pancreatic enzymes with every meal and snack.

“I used to be quite embarrassed about taking Creon at school, people wonder what you are doing...I’ve got used to it though”

“I don’t like taking Creon in front of others, if I go out at night I usually take the Creon before I go out. I don’t like having to explain...people see you taking tablets at lunchtime and ask you questions about what they’re for. Sometimes I take them in the toilet before lunch at school so people don’t see me taking them” (1,4,7,8)

“People think I have problems with asthma because of my cough...I don’t but my cough is there all the time. It was a bit awkward and embarrassing at school” (5)

Six individuals described feelings of frustration regarding the constant need for treatments for their condition. The time consuming nature of physiotherapy and its impact on social activities was a particular issue for respondents.

“I don’t really feel down but I get fed up at times about having to do everything...the constant treatments...it takes so long” (4)

“Having to get physio and take tablets every day can be annoying. It uses up a lot of your time when you could be out playing”

“I still go out with my friends but I have to make time to do my treatments. You need a lot of time to do everything. If I want to go out with my friends I have to be back home by 7pm to fit everything in before bedtime...that’s too early”

“It can be annoying if I want to stay out for longer but I know it (treatment) takes about two hours to do in total so I need to make the time before bed” (1,2,3,8)

“The frequent chest infections and breathing difficulties are frustrating especially if I want to go out but I’m coughing all the time, it’s annoying. I’ve been in hospital a lot recently so I can’t think about work at the moment. It gets really frustrating sitting

around here all day. It used to annoy me if I was doing PE at school and got breathless or tired really easily” (7)

One respondent also commented on the frustration at the unpredictability of the condition

“Sometimes I do everything I’m meant to and still get ill, that’s frustrating” (2)

Importance of Support of Significant Others

All respondents commented on the importance of emotional support from their parents in helping them to manage their condition. Five individuals also mentioned the importance of having supportive friends.

“I have a very close family, they are very supportive of me...they want me to get on with my life as best as I can”

“If plans change at last minute because I’m ill, that’s just the way it is...everyone makes allowances...we cope with the CF together”

“My family, fiancé and his family are all very supportive about my CF...you depend on your family at times” (1,2,3,4,5,6,7,8)

“A lot of my friends know about it (CF) and are very good about it...especially my friends from primary school”

“My close friends are caring and understanding, it makes it easier to cope” (2,4,5,6,7)

Six respondents commented on the importance of practical support from parents to help them to undertake the required treatments, particularly physiotherapy. One individual spoke of the assistance she received from her friends.

“Mum and dad help me a lot...they take turns to do my physio with me” (1,2,4,5,6,7)

“I have three really close friends...they come to stay and help me with my physio and setting up my overnight feeds” (2)

Importance of Acceptance of the Condition and Maintaining a Sense of Normality

Five respondents commented on the importance of acceptance of their condition in maintaining their emotional well-being.

“I feel normal, obviously its not normal but to me it is...I have not known any different...I’ve had it (CF) all my life...my treatment is part of normal life to me”

“I’m active and healthy and CF does not really affect my lifestyle at the moment. I just get on with it...I have no choice. I don’t let CF get me down...I’m not going to let it ruin my life. I do the best I can”

“Its just a habit now. It did affect me a lot after my sister died but as I have got older I have just got on with it. I’ve had it for so long” (2,3,4,5,6)

Six respondents commented on the importance of maintaining a sense of normality to enable them to cope with living with a chronic condition.

“I have to get home IV antibiotics every few months but we try to make everything as normal as possible so I can get on with my life. I was in a school play the last time I had IV’s and just fitted them around the play”

“We take my CF as part of everyday living...I’ve never really missed out on things”

“I still go out with my friends and go away for weekends but I have to make time to do my treatments. When I go for a sleepover I’m allowed the night off from my overnight feed...I usually have my physio and nebulizer before I go and then have it a little later in the morning when I get back home the next day” (1,2,4,5,6,8)

3.4.4.3 Adherence Issues

Reasons for Adherence

Four respondents reported that they adhered to their treatments since they considered them to be important in maintaining their health. One individual spoke of the negative effects of non-adherence to the pancreatic enzymes which encouraged them to remember to take the enzymes in future. Two adolescents commented that parental reminders were the main reasons for their adherence to treatments. In addition, five respondents emphasised the importance of establishing a routine to ensure that the complex treatment regimen required for the condition was undertaken.

“I have always taken the tablets and done my physio pretty much everyday...I know I will get a lot worse if I don’t... my cough gets more frequent and I get tired”

“I do all my treatments to make sure that I stay healthy” (2,4,5,6)

“Sometimes I forget to take my tablets but I suffer the next day. If I’m at a party and have some ‘nibbles’ I won’t think to take my Creon as I don’t think I’m eating much but then I’ll need the next few days off work as I have bad stomach ache” (6)

“My mum and dad remind me all the time to do my treatment” (1,3)

“We have a routine to get everything done, that helps. It’d be difficult to fit everything in otherwise...I don’t know if I’d do it otherwise”

“Treatment is part of my daily routine now. I don’t think about it, I just do it”

“You get used to taking the tablets whenever you get up or with meals, you do it automatically without thinking” (1,2,4,5,7)

Reasons for Non-adherence

The treatment in which motivation appeared to be lowest for the majority of respondents was physiotherapy due to the time-consuming nature of the task which impacted on their lifestyle and the lack of any immediate benefit.

“If I am too tired at night I sometimes can’t be bothered with physio”

“Doing physio everyday can be annoying. It uses up a lot of your time” (1,2,3,7,8)

“I’m meant to do physio and take my nebulizers daily but I don’t really do them...it hasn’t made any difference to my condition...I do physio though if I am feeling unwell” (6,7)

Five of the respondents reported that their parents encouraged and assisted them to undertake their physiotherapy even though four of the respondents had previously reported low motivation regarding this regimen task.

“My parents take turns to do my physio with me, I don’t know if I would do it otherwise” (1,2,3,6,7)

Two respondents reported that low mood had an impact on their adherence to treatments.

“When you feel down you don’t feel like doing anything including treatments” (2,7)

Forgetfulness was also reported by three respondents as a reason for non-adherence to treatments, particularly medication.

“I’m meant to take my tablets four times a day and don’t always take the ones during the day because if I am out I can’t be bothered going back home for them or I forget them” (2,6,7)

No respondents referred specifically to a period of rebellion during their teens which was associated with a lack of adherence to treatment.

Most Difficult Treatments to Adhere to

The majority of respondents reported that the most difficult treatments to adhere to were physiotherapy and intravenous antibiotics.

“I find coughing stuff up and spitting it out during physio difficult, it’s not nice” (1,2,3,6)

“When I get IV’s I do get fed up at times and feel down. You can’t stay with friends or have a shower or dress easily. IV’s are more invasive...you depend on your family and friends at these times to pick you back up when you feel low”

“Home IV’s, that can be a hard time as I can’t go out...it’s harder to cope then...I get so tired but it keeps you healthy” (2,4,6,7)

3.4.4.4 Relationships with Health Care Providers

Satisfaction with the CF Team

There was generally a high level of satisfaction with the CF teams. The majority of respondents reported finding the members of the team supportive on both an emotional and practical level.

“I feel at ease with them (CF team)...had CF all my life so I’ve known them for ages. They are very supportive”

“I still go to the paediatric clinic, the doctor makes me feel listened to. He has got children who are the same age as me and knows how to speak to children and explain things so that I understand them, the language he uses is good”

“I’ve had a close relationship with my nurse ever since my sister died. She has really helped me and I know I can contact her whenever I want. She is more like a friend...if you can’t speak to your family about something then the nurses are always there at the end of the phone. Some things like pregnancy are difficult to discuss with your family but it’s easy to speak to the nurses” (1,2,4,5,6,8)

“Most of the time the nurses can sort out any queries I have even if it is a small one”

“If I ask questions then the nurse will try and find out as much information as possible and relay the information back to me...they support you really well”

(2,4,5,6,7)

Negative Experiences with Health Care Providers

Two respondents expressed a desire for their CF nurse to spend more time getting to know them as a person, believing that at times the nurse was more concerned with getting their job done than trying to form a relationship with them.

“Its good when someone treats me as a person as opposed to someone with CF and tries to get to know me a bit too. The nurse sees me as my CF first and then as me...she is good at her job but it's a bit less personal. It would be nice if she was a bit more friendly” (2,3)

Three respondents described instances of conflict with their consultants regarding their treatment. One respondent reported that they did not always understand the language used within the adult CF clinic. Generally, however, respondents were satisfied with the services received.

“I was ill...the doctor at the adult clinic thought I was not taking the medication to combat the infection and sort of accused me of not taking it when I was...I felt annoyed and quite upset that he didn't believe me” (2,8)

“I don't think that the tablets are helping me so I don't take them now...the doctor keeps insisting that I take them...there's a bit of a battle going on about it at the moment” (6)

“I had to go to the adult clinic for a while because I colonized *Burkholderia cepacia*...sometimes the doctor said things that I didn't understand. The doctor at the paediatric clinic explains things so that I understand them though...the language he uses is good ” (2)

Importance of Autonomy in Treatment

Five respondents commented on the importance of being kept informed about their condition and being included in discussions regarding changes to their treatment

regimens. A sense of autonomy within clinic settings was considered to be extremely important to the young adults in particular.

“They tell me why everything is being done and I’m kept well informed...I do get choices about some things like whether to get the peg tube fitted”

“I usually feel that I can get my point across with the team, definitely can with the nurses”

“I started going to see the doctor myself at the adult clinic...I felt more like an adult. I get a say in my treatments...they ask for your opinion and get your feedback on whether or not you think the treatment is working”

“I am given a say in my treatment, the doctors and nurses always ask if you are happy for changes in doses etc...they are very good at telling you all you need to know about the treatments...I feel included”

“It’s better in the adult clinic because when I was 15/16 years old I got told what was best for me. Now they ask you what you think is best which is a good part of it...you get more of a say...they ask for your opinion”

(1,2,4,6,7)

Four respondents spoke of the difficulties that their parents had encountered regarding the transfer of responsibility for treatment to the individual following the transition to adult CF services.

“Mum finds it difficult as she liked to know what was going on”

“My mum found it difficult not being so involved...she still asks me what the doctor said after each clinic and I fill her in”

“Mum used to do all my treatments with me...she doesn’t see me do it now but she checks the pills in the cupboard are going down I think”

(4,5,6,7)

Confidence in Undertaking Treatments

Five of the respondents reported feeling confident in their ability to undertake their treatment regimens. Two individuals mentioned that the CF nurses had spoken with them at length about undertaking various regimen components which had helped to increase their belief in their competence to carry out the tasks.

“I feel confident about carrying out all the parts of my treatment” (2,4,5,6,8)

“The nurses made sure that my parents and I knew how to administer the overnight feeds, that was helpful...we were more confident about doing it” (2,8)

CHAPTER 4: DISCUSSION

4.1 GENERAL SUMMARY OF RESEARCH

The principal aim of the current study was to apply the self-determination theory of human motivation to explore the relationship between health care professionals' interpersonal styles and adolescents and young adults' adherence to CF treatment regimens. Although the self-determination theory has been applied within other chronic health conditions, there has been no research to date exploring these issues within the CF population. Since less than optimal adherence has been reported in CF patients with significant health implications, information on factors that encourage adherence to regimen tasks is of great importance. The current study also aimed to examine the personal meaning of having CF in adolescents and young adults and includes information on eight individuals who attended for an interview to discuss their experiences in detail. Information was obtained from the interviews regarding the impact of CF on the participants' lifestyles, their emotional reactions to the condition, reasons for adherence and non-adherence to treatment regimens and preferred health care provider relationship styles.

4.2 SUMMARY OF DEMOGRAPHIC DATA

The current study replicates previous literature by Helton, Harmon and Robinson (1991) suggesting that most individuals with CF are diagnosed within the first 15 months following birth, with 73.3% of participants having been diagnosed with CF within the six months following birth. In addition, Shepherd, Hovell and Harwood (1990) reported that adults with CF who were not married or co-habiting were likely to live alone or with their parents compared to their healthy peers who were likely to live with their friends or flatmates. Although the majority of the participants within the current study were adolescents, all of whom were living with their parents, the

study found that of the young adults with CF, three were still living in the parental home with two individuals co-habiting and one participant living alone. Conway (1998) claims that many young adults with CF find it difficult to become fully independent from their parents, on whom they have been dependent for many years for assistance with aspects of their treatment. During the semi-structured interviews two of the young adults who still lived at home reported no immediate plans to move away from the parental home in view of the emotional and practical support they received from their parents and this is in line with Conway's findings. Furthermore, Ricker, Delamater and Hsu (1998) investigated correlates of regimen adherence in children with CF and their families and stated that there was no relationship between adherence and gender, socio-economic status and severity of illness. Within the current study, an Independent t-test and Pearson's Correlations were undertaken to ascertain whether there was a relationship between these variables. No significant results were obtained thereby agreeing with the findings reported by Ricker et al.

4.3 DISCUSSION OF THE HYPOTHESES FROM THE LITERATURE

4.3.1 Hypotheses 1(a), (b) and (c): There will be an association between health care providers perceived as autonomy supportive and participants' autonomous motivation to adhere and increased self-efficacy.

Previous longitudinal studies have examined the influence of the self-determination model on adherence behaviours in a variety of chronic health conditions. Self-determination theory posits that when health care providers are perceived as autonomy supportive, patients display increased autonomous motivation to behave in a particular manner. Furthermore, patients feel more competent to carry out a variety of health related behaviours, resulting in increased adherence to treatment regimens

and improved health outcomes (Deci et al, 1994). In view of these research findings it was predicted that adolescents and young adults' with CF who perceived their health care provider as being autonomy supportive would demonstrate greater autonomous motivation to adhere to their treatment regimens. A bivariate correlation was undertaken and the results supported the hypothesis since there was a meaningful positive correlation at $p < 0.05$ between patients' perceptions of autonomy support from health care providers and their autonomous motivation. In particular, participants' perceptions of autonomy supportive health care providers were meaningfully correlated with their beliefs that looking after their CF was the most important reason for them remaining healthy ($r = .566$; $n=15$; $p < 0.05$) and that effectively looking after their CF was one of the many important aspects of their life ($r = .678$; $n=15$; $p < 0.01$). In view of research by Williams et al (1998b) that self-efficacy is enhanced within the context of an autonomy supportive relationship, it was also hypothesised that adolescents and young adults with CF would display greater self-efficacy if they perceived their health care providers as autonomy supportive. A bivariate correlation was undertaken on these variables and the results indicated that there was a meaningful positive correlation between health care providers perceived as autonomy supportive and greater participants' self-efficacy ($r = .689$; $n=15$; $p < 0.01$). In particular, participants' perceptions of autonomy supportive health care providers were correlated with their feelings that they were able to meet the challenge of looking after their CF ($r = .752$; $n=15$; $p < 0.01$) and also their feelings of confidence in their ability to look after their CF ($r = .635$; $n=15$; $p < 0.01$).

Furthermore, Williams et al (1998a) reported that autonomous motivation was related to a significant increase in diabetic patients' belief in their competence to regulate their glucose levels. A bivariate correlation was therefore conducted to examine whether autonomous motivation to adhere to CF treatment regimens was associated with greater self-efficacy in participants. The results suggested that there was a meaningful relationship between participants' autonomous motivation to adhere and their self-efficacy ($r = .478$; $n=15$; $p<0.05$).

The aforementioned results indicate the importance of an autonomy supportive relationship between health care providers and CF patients, since autonomy support was positively associated with both greater autonomous motivation to adhere and self-efficacy in the participants. Within CF clinics, health care providers are required to educate their patients about the condition and its management and inform them of the importance of consistent adherence to regimens for future health outcomes. The results of the current study would suggest that it is not sufficient solely to provide patients with this information. Health care providers are required to adapt information depending on the cognitive developmental level of the patient in order to ensure understanding and also to consider the implications of the condition and its treatment on the individual's lifestyle. In addition, self-determination theory emphasises that patients should be provided with an opportunity to ask questions and voice concerns regarding the proposed regimens since individuals will display increased interest in particular behaviours being supported by health providers, such as the importance of consistent adherence to regimens, if they perceive themselves to be involved in the decisions about their self-care (Williams, Frankel, Campbell and Deci, 2000).

Moreover, Williams et al (2000) state that providers perceived as controlling will cause patients to feel coerced to behave in a particular manner and Williams et al (1999) report that a controlling relationship may also increase anxiety levels within the individual, with any behaviour change likely to be short lived and poorly integrated into the individual's long-term values. An additional bivariate correlation was therefore conducted to ascertain whether perceptions of autonomy supportive health care providers were related to controlled motivation to adhere. As would be predicted from the self-determination model, no significant correlation was found between autonomy supportive health care provider relationship styles and participants controlled motivation ($r = .206$; $n=15$; $p = .231$). Autonomy support was not related to participants' feeling pressurised to behave in a certain manner and the influence of autonomy support on patients' motivation to adhere is therefore an important aspect for health care providers within CF teams to consider. An additional correlation was conducted between scores on the measure of self-efficacy and participants' controlled motivation to adhere. As would be anticipated, no significant correlation was found between self-efficacy and participants' controlled motivation ($r = .294$; $n=15$; $p = .144$). One of the most interesting findings of the current study was the significant relationship between providers perceived as autonomy supportive and participants' self-efficacy. It would therefore appear to be important for health care providers to develop an understanding of patient's beliefs in their own competence to carry out treatment regimens in addition to providing them with choices and a say in decisions regarding the management of their condition in order to encourage more consistent adherence to regimens.

4.3.2 Hypothesis 1(d): Adolescents and young adults who report greater self-efficacy will display increased adherence to CF treatment regimens.

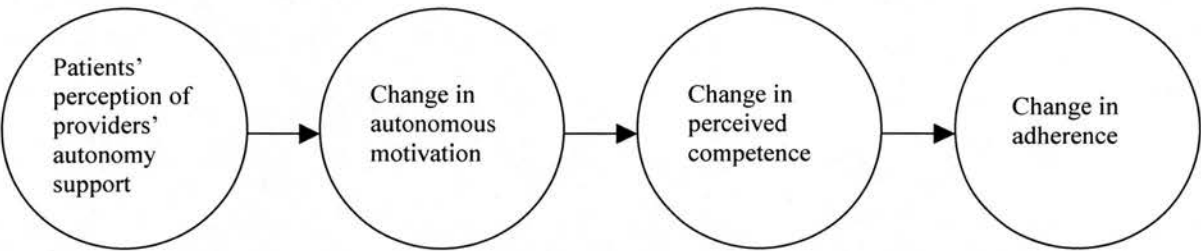
In view of findings by O'Leary (1992), Bartholomew et al (1993) and Williams et al (1998a) that self-efficacy has an influential effect on adherence behaviours, it was anticipated that there would be a correlation between increased self-efficacy and greater adherence to treatment regimens. The results of the current study indicated that there was a meaningful relationship between participants' self-efficacy and their adherence to diet ($r = .466$; $n=15$; $p<0.05$), although no significant correlations were found between participants' self-efficacy and their adherence to medication, physiotherapy or exercise.

The majority of respondents who attended the semi-structured interviews commented that the first regimen task that they were given responsibility for undertaking was the consumption of pancreatic enzymes within the school environment and they reported feeling competent to manage this task. Furthermore, they reported that non-adherence to pancreatic enzymes was associated with immediate negative symptoms of abdominal pain that consequently encouraged them to take their enzyme replacements. It may be that parents were more influential in the individual's adherence to physiotherapy by way of frequent reminders and assistance to undertake the regimen task. In addition, parents may look after medication and tablets required by children and adolescents and administer them when required. Furthermore, exercise is generally included as part of the school curriculum. Adherence to these three treatment components, particularly during adolescence, are therefore unlikely to be influenced by whether or not an individual feels competent to carry them out thereby accounting for the non-significant results obtained. A larger sample size is

required to confirm this result, since a post-hoc power calculation indicated that there was only a 20%, 16% and 5% (respectively) chance of detecting a significant relationship between the variables with the current sample.

The self-determination model developed by Williams et al (1998a) is illustrated in Figure 6 below and shows that patients' perceptions of autonomy support from health care providers results in increased autonomous motivation to adhere in diabetic patients. This in turn results in an increase in perceived competence and consequently better adherence.

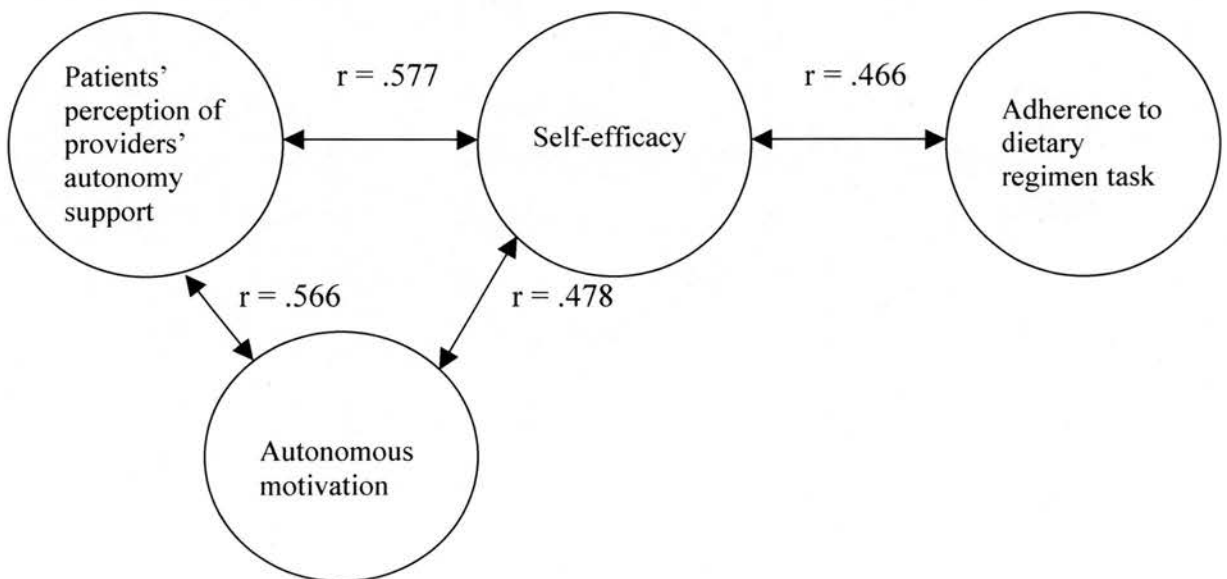
Figure 6: The self-determination model (Williams, Freedman and Deci, 1998a)



The results of the current, investigative study provide some evidence that the self-determination theory could be applicable within the CF population since meaningful correlations were obtained between the factors inherent within the theory. Although correlations do not necessarily imply causality, this study demonstrates that there are particular relationships between the perception of autonomy supportive health care providers and participants' autonomous motivation to adhere and their self-efficacy. Furthermore, the results suggest a relationship between participants' self-efficacy and their adherence to the dietary regimen task.

Whilst controlling for autonomous motivation to adhere, a partial correlation was conducted between autonomy support and participants' self-efficacy to establish whether autonomous motivation was a mediating variable between autonomy support and self-efficacy. The partial correlation suggested that autonomous motivation was a mediator in the link between autonomy support and self-efficacy, as suggested by the self-determination model, since the correlation between these two variables was less significant when controlling for participants' autonomous motivation ($r = .577$; $df = 12$; $p < 0.05$). The finding suggests that autonomy support had an impact on participants' self-efficacy regardless of whether the participant was motivated to adhere, although autonomous motivation is a mediating variable. A diagram of the results of the present study is displayed in Figure 7.

Figure 7: Diagram depicting the results of the current study



The model derived from the current study suggests that while autonomous motivation is a mediating factor between autonomy support and self-efficacy, autonomy support does appear to be directly associated with self-efficacy

regardless of whether an individual is autonomously motivated to adhere. The cross sectional nature of the study does not allow for causal interpretation of the results and further studies of a longitudinal nature are required to investigate the causal direction of the association between autonomy support, autonomous motivation and self-efficacy. In addition, multiple regression analysis may have proved useful to establish the relative importance of autonomy supportive relationship styles, autonomous motivation and self-efficacy in predicting adherence to treatment regimens. However, due to limited numbers of participants in this study a multiple regression analysis was not conducted, although with a larger sample such detailed analysis of the data may have yielded interesting results.

4.3.3 Hypothesis 2: Lower levels of psychological distress (anxiety and depression) will be reported by adolescents and young adults with CF who perceive their health care providers as autonomy supportive.

It was predicated that those participants who perceived their health care providers as being autonomy supportive would demonstrate lower levels of psychological distress. Williams et al (1999) undertook a longitudinal study of medical students taking an interview course and suggested that when instructors were perceived as autonomy supportive, the students displayed greater conceptual learning and better psychological adjustment in terms of lower anxiety, higher self-esteem and more positive affect. No significant correlations were found between perceptions of autonomy supportive health care providers and levels of anxiety and depression in the current study, although the results suggested a trend in the anticipated direction with lower levels of anxiety and depression being experienced if health care providers were perceived as autonomy supportive. Williams and colleagues claimed

that autonomy support of instructors promoted the active engagement and a sense of volition with respect to learning in the students who participated in their study. A larger sample size may have shown more meaningful correlations between the presence of autonomy supportive health care providers and the psychological well-being of participants, with participants who do not feel coerced or pressurised to behave in a particular manner reporting reduced levels of anxiety as suggested by Williams et al. A post-hoc power calculation indicated that for the current study, with a sample size of 15, there was only a 20% chance of detecting any significant result between perceptions of autonomy support and anxiety and a 17% chance of detecting any significant result between perceptions of autonomy support and depression.

4.3.4 Hypothesis 3: Adolescents and young adults with CF who report greater self-efficacy will display lower levels of psychological distress (anxiety and depression).

O'Leary (1992) reported that participants high in self-efficacy were not only more likely to adhere to medical regimens but were more likely to experience less stress and negative emotional states. It was therefore anticipated that participants presenting with high self-efficacy would report lower levels of psychological distress. No significant correlations were found between participants' self-efficacy and their levels of anxiety and depression, although the results suggested a trend in the hypothesised direction with lower levels of anxiety, in particular, being experienced if participants' self-efficacy is high. A larger sample size is required to detect whether there is any significant association between self-efficacy and psychological adjustment since a post-hoc power calculation indicated that there was

only a 36% chance of detecting any significant result between self-efficacy and anxiety and a 13% chance of detecting any significant result between self-efficacy and depression.

4.4 DISCUSSION OF THE EXPERIMENTAL HYPOTHESES

4.4.1 Hypothesis 4: Higher levels of psychological distress (anxiety and depression) in adolescents and young adults with CF will be associated with poorer adherence to CF treatment regimens.

It was anticipated that psychological distress would impact negatively on adherence to treatment regimens. However, no significant correlations were found between participants' levels of anxiety and depression and their adherence to the individual CF treatment regimen tasks. Nevertheless, the results suggested that higher levels of anxiety and depression had a possible negative impact on adherence to diet in particular ($r = -.168$; $n=15$; $p = .275$ for anxiety and $r = -.361$; $n=15$; $p = .093$ for depression). Although the results indicate a weak relationship, this finding is in line with previous literature which has reported on the negative effect of psychological distress on adherence (Koocher et al,1990). Furthermore, this finding would be expected due to the acknowledged impact of anxiety and depression on appetite. Gilbert (2000) reports that as a result of the physical changes that occur within the body during episodes of depression and anxiety, not only are energy levels and sleep affected but a loss of appetite is also common.

One reason for finding no significant association between psychological distress and reduced adherence to medical treatment, physiotherapy and exercise may be due to the fact that the majority of participants in the sample fell within the 'normal' range

on the Hospital Anxiety and Depression Scale, with only three participants demonstrating clinical 'caseness' for anxiety and no participants presenting with clinical 'caseness' for depression. The sample group therefore did not present as having particular difficulties with psychological distress thereby reducing the likelihood that any association would be found. Furthermore, the qualitative data obtained from the interviews suggests that, although the individual may be experiencing problems relating to their emotional well-being, the emotional and practical support from significant others, in the form of reminders and assistance to adhere to their regimen tasks, may prevent the individual from neglecting their regimens, thereby validating the non-significant results obtained in this study.

In addition, within the diabetes literature, Anderson, Nowacek and Richards (1988) stated that the personal meaning of diabetes amongst adults was strongly related to their self-care and the degree of psycho-social adaptation. The sample group in this study appeared to display high levels of psycho-social adaptation to their illness and this may explain the lack of any significant findings. The hypothesis was therefore not supported. Consequently, a post-hoc power calculation was conducted which indicated that there was approximately an 8% chance of finding any significant relationship between reduced adherence to the dietary regimen task and levels of anxiety and approximately a 27% chance of finding any actual relationship between reduced adherence to the same regimen task and levels of depression. A larger sample would therefore be required in future studies.

4.4.2 Hypothesis 5: Increased age will be associated with better adherence to CF treatment regimens.

It was predicted that increased age would be associated with better adherence, in view of the substantial literature emphasising difficulties with adherence in adolescents as a result of their search for independence and desire for acceptance by peers which can outweigh the demands of a self-care regimen (Cameron and Gregor, 1987; Tyrrell, 2001). No significant positive correlation was found between increased age and better adherence to the four regimen components examined in this study and this hypothesis was therefore not supported. Nevertheless, the results did suggest a meaningful negative correlation between adherence for physiotherapy and increased age ($r = -.491$; $n=15$; $p<0.05$). This result could be attributed to the fact that many CF patients require assistance with physiotherapy from their parents until early adulthood (Conway, 1998). The qualitative data also indicated that many of the respondents were regularly reminded and assisted to adhere to this specific regimen task by their parents. Furthermore, the respondents who attended the interviews commented on the importance of establishing a routine for their treatments, particularly physiotherapy due to its time-consuming nature. Parental reminders to adhere and the presence of a routine may have increased the likelihood of physiotherapy being undertaken by the adolescents within the sample, particularly since they were all living within the parental home whereas three of the young adults in the sample were now living outwith the parental home and would therefore have received less reminders and assistance from their parents to do their physiotherapy on a daily basis. In addition, young adults who are trying to maintain a normal lifestyle may decide not to carry out physiotherapy because of the time required to undertake the task and the lack of immediate perceived benefits (D'Angelo and

Lask, 2001). Due to the small sample size of the study and the lack of any significant correlations, a post-hoc power calculation was conducted and indicated that there was approximately a 12% chance of finding a significant relationship between increased age and adherence to exercise and approximately a 26% chance of finding an actual relationship between increased age and adherence to the dietary aspects of the regimen. A larger sample would be required in future studies in order to ascertain whether there is indeed any association between increased age and adherence to medication, diet and exercise regimens.

4.4.3 Hypothesis 6: There will be an association between increased severity of CF and increased psychological distress (anxiety and depression).

It was anticipated that as participants' health deteriorated, as indicated by a reduction in their FEV1 % predicted, their levels of anxiety and depression would increase. However, no significant correlation was found between severity of the condition and levels of anxiety or depression in adolescents and young adults with CF. Nevertheless, the results indicated a trend in the anticipated direction for anxiety in particular, with increased anxiety being experienced with any deterioration in their condition ($r = -.336$; $n=15$; $p = .111$).

There may be several reasons for the lack of an association between a deterioration in health and levels of anxiety and depression. Respiratory failure is responsible for 97% of all CF deaths (Penketh et al, 1987) and lung transplantation is suggested if an individual presents with a severe deterioration in lung function, as indicated by a FEV1 of less than 30% of their predicted value, and a significant reduction in quality of life (Dark and Corris, 1996). The suggestion that a patient requires lung

transplantation can be highly stressful for patients and their families, since it implies a serious risk of death. However, within the sample group an average FEV1 % predicted value was calculated for the previous six months. The mean FEV1 % predicted reading was 77.33 (SD=13.96) with a range of 52-99 thereby indicating that none of the participants had an FEV1 % predicted reading near to the cut-off implying the need for lung transplantation. The qualitative data also suggested that all participants appeared to be functioning well within their daily life despite having CF and this may partly explain the lack of any reported problems with anxiety and depression. FEV1 % predicted can also be affected by respiratory infections and consequently, even if an individual's FEV1% predicted was lower than usual, the reduced reading might be attributable to a chest infection, which is often treated by antibiotics or an increase in time spent on physiotherapy, as opposed to signifying a deterioration in health and therefore not cause significant distress to the individual.

Furthermore, emotional reactions to any deterioration in health may be cushioned by the level of emotional support available from significant others. The participants interviewed indicated that they received regular input from the CF teams and felt confident that any deterioration in health would be competently managed by the team. In addition, Shepherd et al (1990) report that CF sufferers have a tendency to perceive themselves as 'less severe' than their peers with CF. During the semi-structured interviews, three respondents commented on this fact: "I'm more healthy than lots of other people with CF I know". It may be that the individuals within this study have adapted to their condition well, are well supported and are therefore more able to cope with any deterioration in health. Although no measure of coping style was utilised in this study, it may be interesting in future research to investigate

whether particular coping styles have any impact on the level of psychological distress experienced by CF patients during a deterioration in their condition. An additional reason for the lack of any significant results may be due to the measure of severity of illness utilised. The use of a clinical rating of severity such as the Shwachman score (Shwachman, 1965) may have demonstrated different results. The Shwachman score classifies patients into severe, moderate, mild, good and excellent depending on their total score and the rating is based on the sum of scores across four parameters: general activity, physical examination, nutrition and chest x-ray. Many CF services complete this clinical rating of severity at each clinic appointment, however, the Tayside and Forth Valley clinics did not consistently utilise the Shwachman score and data was therefore unavailable for the majority of the participants.

Moreover, Sensky (1990) reports that patients' reactions to chronic illnesses often depends less upon the severity of the illness than upon the patients' attitude towards the illness. Within this sample group the participants appear to have adjusted well to their illness, have been encouraged to lead as normal a life as possible despite their condition and have been well supported by their families and the CF team. This may therefore have positively impacted on their ability to manage any deterioration in health and may explain the findings. The hypothesis was not supported and various possible reasons for this have been discussed. However, the sample size of this study was small and a post-hoc power calculation suggested that there was approximately a 16% chance of finding a significant relationship between severity of illness and anxiety levels. Furthermore, there was approximately a 10% chance of detecting an actual relationship between severity of illness and levels of depression. A larger

sample would therefore be required in future studies in order to ascertain whether there is indeed any association between increased severity of illness and psychological distress.

4.5 DISCUSSION OF THE QUALITATIVE DATA

The aim of the qualitative analysis was to report the diversity of experiences amongst adolescents and young adults with CF in line with the hypotheses of the study. In particular, the impact that the condition had on their lifestyle, their emotional reactions to their condition, reasons for adherence and non-adherence to regimens and their relationships with health care providers. Due to the small numbers involved in this part of the current study, no claim is made that the opinions expressed represent the experiences of young people with CF in general. The most frequently reported themes during the interviews concerned the importance of acceptance of the condition and attempting to maintain a sense of normality to lessen the impact of the condition on the individuals' lifestyles. In addition, establishing a routine for the treatment regimens and emotional and practical support from parents, friends and health care providers were considered to be extremely valuable and appeared to be interrelated to adherence to regimens. The majority of respondents also described a preference for a sense of autonomy within health care settings when discussing treatment regimens. From the information gathered from the respondents during the interviews, it is evident that within this sample group, having CF was perceived as something that they had to learn to accept and live with. Contrary to expectations, the majority of respondents did not report significant difficulties with anxiety or low mood although the time-consuming nature of treatment regimens, particularly physiotherapy, was a major cause of frustration for some in their lives.

4.5.1 Discussion of the Qualitative Findings

An examination of the qualitative data indicated that having a chronic illness disrupted school attendance due to prolonged absences because of poor health and frequent hospital admissions, which in turn interfered with the participants' academic work and limited their level of involvement with their peers. The majority of participants, however, reported that their schools had been accommodating with regard to their absences and had attempted to assist them to keep up to date with their class work. They also reported that they had not been segregated or bullied at school or made to feel particularly different from their peers. In relation to hospital admissions for intravenous antibiotics or due to ill health, several of the respondents reported missing their families whilst in hospital but generally found their time in hospital to be tolerable. One respondent commented on feelings of isolation during hospital admissions since they had to be segregated from other CF patients because they had colonized *Burkholderia cepacia*. The respondent stated that they had previously mixed with their CF peers and had discussed illness-related thoughts, concerns and strategies regarding how to manage the condition with them. Consequently, they had found their recent segregation difficult. However, the respondent commented that they were still able to communicate with their peers via e-mail which had been useful. The presence of *Burkholderia cepacia* in CF patients has severely curtailed their opportunity for mutual support from other CF patients due to the risk of cross infection for peers (LiPuma, Dasen, Nielson, Stern and Stull, 1990). New technology such as e-mail and internet support groups may therefore prove valuable to these individuals to enable them to maintain communication whilst avoiding face to face contact and prevent them from feeling as isolated.

With regard to emotional well-being, five respondents spoke of the importance of acceptance of their condition in maintaining their emotional well-being. Furthermore, acceptance of the condition appeared to be linked to the importance of maintaining a sense of normality. Six respondents considered this to be essential in dealing with their CF. Acceptance of the condition and attempts to live as normal a life as possible despite having CF were common strategies used by respondents in this group. With the prospect of having a life-long condition with an unpredictable and limited life expectancy, these are perhaps effective strategies for dealing with CF. These factors also appeared to be related to reports by the majority of respondents that they did not think about their future health in detail and instead tried to get on with and enjoy their life as much as possible.

The majority of respondents also spoke of the importance of emotional and practical support from significant others, including parents and health care providers, in assisting them to manage their condition. This support was also perceived as valuable in encouraging them to adhere to treatment. The qualitative data also suggested that some of the respondents appeared to cope better with having CF than others as indicated by the range of emotional reactions reported during the interviews. Emotional reactions most frequently described included embarrassment about having to take pancreatic enzymes within social situations, particularly within the school environment, as this made participants feel different to their peers. Frustration at the time-consuming nature of physiotherapy, the impact of intravenous antibiotics on social activities and the constant need for treatment was also reported by several respondents.

Having a chronic condition, like CF, can be very stressful for the whole family as the mornings and evenings, in particular, are governed by treatments. Five respondents described the importance of establishing a routine for treatments to allow them to balance a normal life with their need for treatment. In relation to reasons for adherence, four respondents commented on the importance of adhering to treatment in order to maintain their health. The majority of respondents described feeling confident in undertaking their treatment and reported that they had been provided with sufficient information about treatments from their health care providers. Moreover, although five respondents described low motivation to undertake physiotherapy in particular, due to its time-consuming nature and the lack of any immediate benefit, four of these individuals stated that their parents often encouraged them to adhere with this regimen component and provided assistance with it.

The majority of respondents who attended the interviews generally perceived their care providers to be autonomy supportive. They described being given choices about their treatments and feeling that their viewpoints were understood and considered themselves to have some autonomy with regard to decisions about their regimens, particularly within the adult CF clinics. Respondents appeared to be satisfied with the services received from the CF team care and many found the CF liaison nurses particularly easy to talk to about a range of matters.

In general, the participants who attended the interviews seemed to have had positive experiences with their health care providers and described supportive relationships with their families and peers. Comments obtained from the interviews would suggest that this support was important in promoting respondents' beliefs in their ability to

adhere to their treatment regimens. These findings agree with Eiser (1993) who stated that how a child adjusts to their illness is related to their health and their family and social environment. Those participants interviewed perceived CF as part of their life and treatment regimens generally appeared to be accommodated into their lifestyle. It is possible that the individuals who declined to be interviewed, or indeed participate in the study, may have given a different perspective of their adaptation to the condition, their level of support from family, peers and health care providers and their subsequent adherence to regimens.

Clearly not all the comments made during the interviews could be categorised into the main themes given above. Those that have been identified demonstrate the extent to which adolescents and young adults with CF deal with their condition and describe factors associated with assisting the respondents to maintain self-management. Individuals presenting with emotional difficulties in relation to their condition may require psychological input to enhance their sense of control over the condition and effectively manage the associated stresses (Hains et al, 1997). However, as mentioned in the introduction, not all CF teams have a clinical psychologist within their membership. The need for psychological input to be available for young people with CF and the possible inclusion of a clinical psychologist within all CF teams is an area that requires further investigation. Certainly discussions with the consultant paediatricians and consultant respiratory physicians indicate a desire for resources to be made available to enable this.

4.5.2 Methodological Issues Relating to the Qualitative Data

It should be taken into consideration that the interpretation of the qualitative data in the current study did not strictly adhere to any specific qualitative approach. The coding of the transcript information could have been improved by utilising a more formal method of identifying themes from the data such as grounded theory, developed by Glaser and Strauss (1967). However, this was not possible due to the time limitations of the study. In addition, an attempt was made to link qualitative and quantitative concepts and therefore the interpretation of the information obtained from the semi-structured interviews may have been compromised as themes were primarily identified from previous research as opposed to being solely derived from the data. Coding could have been further enhanced by a second researcher recoding the transcripts thereby minimising researcher bias. However, investigation of inter-rater reliability was not possible due to the time constraints of the study. The size of the sample for the qualitative analysis also limits the extent to which the results and themes can be generalised to the CF population and therefore the analysis of the transcripts may only be pertinent to this particular sample group. Future research within this area would require a larger sample to determine whether the themes identified within this study were representative of the CF population as a whole. Nevertheless, the use of semi-structured interviews, with open-ended questions, allowed for detailed and interesting information regarding the personal experiences of adolescents and young adults diagnosed with CF which augmented the quantitative results obtained. Although the interviewer directed the structure of the interview to an extent, it is unlikely that this diverse information would have been obtained through the use of questionnaire methods alone.

4.6 GENERAL METHODOLOGICAL ISSUES

With regard to the general methodological issues, there are several points that warrant consideration and have implications for the interpretation of the results and these will now be discussed.

4.6.1 The Recruitment of Participants

Difficulties were encountered in the recruitment of CF patients into the study and for this reason the geographical area from which participants were recruited had to be broadened. The study was well received by the consultant paediatricians and consultant respiratory physicians within the two Health Boards as they considered it to be pertinent to the recognised problems of non-adherence within CF services and valuable in providing information regarding the possible influence of patient-physician interactions to address the problem. However, the number of participants who agreed to participate in the study fell considerably short of the numbers that had initially been anticipated by the consultants, with a disappointing response rate of 22.4% and the consultants expressed surprise at the poor response. There may be several reasons for the low response rate and these will be discussed in turn. In the current study, questionnaires were posted to individuals due to time limitations, however, as Lebow (1982) reports there is a tendency for low response rates associated with studies involving postal questionnaires. In retrospect it may have been beneficial to meet with selected individuals within the CF clinics, with ethical approval, to issue the information sheets, consent forms and questionnaire packs as this may have reduced any anxiety and have increased the likelihood of additional responses. Individuals may also have been concerned about the confidentiality of their responses, particularly as some of the measures used in the study required them

to discuss their relationship with their health care providers. They may therefore have been reluctant to participate, despite assurances of confidentiality of the information received and emphasising that the researcher was not part of the CF team.

Another reason for the poor response rate may be due to the fact that CF requires time-consuming regimens and regular visits to the paediatric and adult CF clinics. The invitation to participate in the study may have been perceived by the CF patients as an additional burden related to their condition, even though the completion of the questionnaire pack was estimated to take approximately 30 minutes in total. Furthermore, CF patients miss school more than their healthy peers due to ill health, hospital admissions and clinic appointments and this can make it difficult for the individual to keep up to date with their academic work (Fowler, Johnson and Atkinson, 1985). At the time of recruitment many adolescents were in the process of sitting examinations at school and therefore the study may have seemed like an additional stressor prior to the examinations. Indeed, four individuals contacted the researcher to state that they were currently revising for examinations and were therefore unwilling to participate in the study.

Moreover, Suls and Fletcher (1985) have described two defence reactions exhibited by individuals with chronic health conditions. The first is an avoidance response, sometimes called 'denial', that is implemented in an attempt to minimise threat by focusing attention away from the condition and consequently the individual may show little interest in information about the disease or changes in the condition. The other defence reaction is to monitor the disease and follow any changes in the disease process closely and Suls and Fletcher state that individuals who implement this

strategy may be more willing to talk openly about their condition. Strauss and Wellisch (1981) found a prominent use of 'adaptive denial' amongst CF sufferers and their families and claimed that although the negative reality of the illness was recognised, patients and their families tried not to think about the condition more than necessary and focused instead on daily activities. It may be that a proportion of the non-responders utilise the avoidance response and were therefore discouraged from participating in the study, as they did not wish to think of or speak about their condition. In contrast, those individuals who participated in the study could possibly be considered to fit the other defence strategy in view of their willingness to discuss their condition and its effect on their lives. Further research investigating the reasons for non-participation in CF-related studies may provide interesting information that could be utilised to encourage future participation in studies.

4.6.2 The Generalisation of the Results

The low response rate to this study limits the conclusions that can be drawn from the research and the extent to which the results can be generalised to the CF population. In order to ascertain whether the individuals who participated in the study were representative of the CF population as a whole, it would have been useful to obtain demographic information on non-respondents. This was not possible, however, in order to safeguard the confidentiality of patients, as the Medical Directors within Tayside and Forth Valley NHS Health Boards granted Caldicott Guardian approval to access the hospital case notes of only those individuals who gave informed consent to participate in the study. Nevertheless, the quantitative and qualitative data obtained was interesting and the results should be considered useful as a pilot study for future research.

4.6.3 The Measure of Adherence Utilised

Rapoff (1999) states that there is no ideal measure of adherence at present, with all techniques having at least some methodological limitations. However, he reports that patient and parental estimates of adherence are regularly used within clinical settings. Within this study, patient self-reports of adherence to four regimen components were obtained. In order to aid recall, adherence was to be rated for the previous week and, as suggested by Lask (1994), adherence behaviour was conceptualised along a continuum. Furthermore, in an attempt to ensure that estimates of adherence were accurate, it was emphasised that there were no right or wrong answers and the confidentiality of responses was assured. Obtaining several reports of adherence behaviour across different time periods may have provided interesting information regarding possible variations of adherence across time, although this was not possible due to time limitations of this study. Moreover, it may have been worthwhile to obtain a provider estimate of adherence which could then be compared with patients' self-reports and any discrepancies between these reports subsequently investigated.

4.6.4 Time Delay Incurred whilst Awaiting Ethical Approval

Research ethics committees (RECs) have been criticised for the time delays incurred by researchers whilst awaiting ethical approval to proceed with research (Cohen, 1999) and such a problem was encountered with this study. A research proposal for the study was submitted to the Tayside Committee on Medical Research Ethics for their committee meeting in December 2001. Approval to proceed was obtained, subject to minor amendments, and the amended documentation was returned to the secretary of the committee at the beginning of January 2002 for review. However,

formal approval was not granted until seven weeks after the amendments were submitted because of annual leave and sick leave of committee members which resulted in a backlog of proposals. The starting date of the research was therefore delayed which was particularly frustrating given the strict time limitations of the study. Furthermore, due to the low response rate in Tayside an additional proposal was submitted to the Forth Valley Ethics of Research Committee in April 2002. Relatively minor amendments were requested, however, the researcher was invited to discuss these amendments with the committee at the next meeting in May. Although formal approval was granted following the meeting it was considered that the issues could have been resolved by written correspondence thereby preventing a further time delay to the study.

The importance of early submission of future proposals to RECs was recognised in view of the possible time delays incurred. However, it is hoped that the guidelines specified within the Department of Health Research Governance Framework, introduced in April 2002, will reduce unnecessary time delays between receiving and reviewing proposals or amendments, as RECs are now required to submit proposals to another REC if delays are anticipated. RECs have also been criticised for the diversity in practice between different Health Boards (Cohen, 1999). In the current study different amendments were requested by the RECs within Tayside and Forth Valley. The implementation of a standard protocol for ethical review, as proposed in the research governance framework, should however resolve this issue for future researchers.

4.6.5 Post-Hoc Power Calculations

In view of results from a similar study by Howells et al (2001) that applied self-determination theory within the asthma population, a large effect size was anticipated. Power calculations suggested that at least 40 participants were required to produce a large effect size for the statistical analysis used and the consultant paediatricians and consultant respiratory physicians were confident that sufficient participants could be recruited. Fewer participants were recruited than was anticipated, however, and consequently post-hoc power calculations have been calculated and discussed within this chapter. A continuation of this study with a larger sample may result in additional, more robust findings.

4.7 CLINICAL IMPLICATIONS OF THE STUDY

4.7.1 Implications of the Study for Health Professionals in CF Teams

Despite the low response rate, the results of the study suggest that there is a meaningful correlation between participants' perceptions of autonomy supportive health care providers, participants' autonomous motivation to adhere to treatment regimens and their self-efficacy. Increased self-efficacy is also associated with better adherence to dietary aspects of the CF regimen. In view of the findings, the results of the current study will be relayed to health care providers within the CF teams within the two Health Boards where the study was conducted and a continuation of the study with a larger sample recommended to confirm the findings.

Within the literature many studies have focused on the impact of patient, family and regimen factors on adherence but relatively few have examined the role of the patient-practitioner interaction. Clark et al (1995) posit that the physician-patient

relationship is especially important in the management of chronic illness as the physician helps the patient assume responsibility for implementing and actively monitoring the therapeutic regimen and guides the patient when adjustments to the regimen are required. The importance of a collaborative, supportive and non-judgemental relationship between patients, family members and health care providers on adherence is also advocated by the self-determination theory of human motivation (Deci and Ryan, 1985).

Williams et al (2000) reviewed the literature concerning health care outcomes associated with autonomy supportive patient care and concluded that when physicians were more autonomy supportive than controlling, patients were likely to display higher satisfaction with health care, better adherence to prescriptions, more maintained behaviour change and better physical and psychological health. In view of the research applying the self-determination theory to chronic conditions and the preliminary results of this investigative study, it would seem appropriate to educate members of the CF teams about the importance of autonomy supportive relationships with patients to enable them to offer optimal support to young people with CF.

Westwood et al (1999) conducted a cross sectional study of adolescents and adults with CF regarding their perspectives about the transition to adult CF clinics and found that autonomy in health care was deemed to be 'extremely important' by 77% of respondents. The qualitative data obtained in this study supports this finding with a sense of autonomy within CF clinics being perceived as important by the majority of respondents. Relationships between adolescents and young adults with CF and their health care providers, where the patient feels a sense of autonomy and is

provided with information to enable them to make informed decisions concerning their adherence to prescribed regimens, appear to have a beneficial effect not only on patients' motivation to adhere but also their confidence in their ability to take responsibility for and carry out their treatments.

4.7.2 Implications for Improving Adherence to Treatment Regimens

Adolescents and young adults with chronic conditions have unique issues and requirements and have to balance treatment regimens with everyday activities. Rapoff (1999) states that attempts should therefore be made, where possible, to negotiate treatment goals and reduce the complexity and intrusiveness of regimens to suit the lifestyle of the individual and thereby improve the likelihood of adherence. During the semi-structured interviews, one respondent commented on the positive impact that negotiations about their regimen had on adherence: "I'm meant to use my nebulizer four times a day but I often forget to or can't be bothered going home during the day to do it...the doctor suggested that I try using it in the morning and at night for twice as long as normal...that's easier to do because I already take my tablets then anyway". The importance of negotiating regimens to consider the lifestyle and social context of patients to maximise adherence will also be discussed with the CF teams.

Within this study, increased self-efficacy appeared to be related to increased adherence to dietary aspects of the CF regimen. Health care providers therefore require to be informed of the importance of enhancing the self-efficacy of CF patients regarding their ability to carry out treatment regimens. Bandura (1997) posits that attempts to enhance self-efficacy are most effective if the provider is

viewed as trustworthy, competent and autonomy supportive. He states that through emphasising their belief that the patient is capable of doing the required tasks and ensuring that the patient is successful in their efforts, health care providers can increase patients' feelings of competence regarding their self-management. Clinicians also need to be sensitive to concerns regarding regimen tasks since open communication can lead to effective problem solving and consequently increase the confidence of the patient to undertake the regimen components. During the semi-structured interviews, two respondents reported that they had been apprehensive about undertaking overnight feeds but had felt more able to carry out this task, with the assistance of their parents, following discussions with the CF nurses regarding their concerns: "The nurses made sure that my parents and I knew how to administer the overnight feeds, that was helpful...we were more confident about doing it".

Czjakowski and Koocher (1987) found that adolescents with CF who believed that their actions made a difference to their health displayed good levels of adherence to their treatment. Kyngas and Barlow (1995) also reported that in addition to increasing knowledge about health conditions and skills required to implement treatments, there is also a need to improve the self-efficacy of the individual to increase their confidence in their ability to carry out their self-care. In view of the findings from the literature and the current study it would appear necessary to monitor patients' self-efficacy within CF clinics and address low self-efficacy in patients since this may impact on their adherence. However, although there have been attempts to develop and validate illness-specific self-efficacy scales for children and adolescents in diabetes and asthma, no such measures are presently available for the CF population. Future research to develop or modify a self-efficacy measure for

this client group would be beneficial. Moreover, patients' belief in the likelihood that their efforts will reap positive benefits also appears to be important for their adherence to regimens. Rapoff (1999) claims that clinicians should emphasise the potential benefits of prescribed regimens, provide disease outcome information and encourage patients to monitor symptoms to demonstrate the benefits of the prescribed regimens.

4.8 FUTURE STUDIES

Throughout this chapter, suggestions for future research in relation to the hypotheses outlined have been expanded upon. Additional areas for future investigation will now be discussed. The qualitative data suggested that within this sample group, adolescents and young adults with CF were relatively well adjusted to their condition in terms of their emotional well-being. Detailed information was obtained from the interview transcripts that indicated that several respondents viewed acceptance of their condition as important in enabling them to maintain a sense of normality. However, no measure of acceptance of illness was used in this study. Future research could consider utilising the Acceptance of Illness Scale (AIS), developed by Felton, Revenson & Hinrichsen (1984), that directly assesses adjustment to illness. If CF patients are perceived as resilient and adaptable to their illness as found in this study and also suggested by Angst (2001), possible reasons for this resilience could then be examined in detail and the results applied to other chronic conditions.

In addition, although the application of the self-determination theory appears to be relevant to CF patients, the conclusions drawn from this study will need to be supported by further research using a larger sample of adolescents and young adults

with CF. A longitudinal study examining the influence of autonomy supportive health care provider relationship styles on patients' self efficacy, motivation to adhere, subsequent adherence behaviours and health outcomes over a 6 and 12 month period would be recommended.

4.9 CONCLUSIONS

Non-adherence to treatment regimens in young people with CF and the subsequent impact on health care resources, school and work absenteeism and long-term health outcomes has been well documented within the literature. It is therefore important that research examines factors likely to encourage maximum adherence within this population. However, psychological research investigating the importance of health care provider relationship styles on CF patients' motivation to adhere, self-efficacy and consequent adherence to treatment regimens is still in its infancy. The results of the current, investigative study suggest that young peoples' perceptions of autonomy supportive health care providers were associated with an increase in their self-efficacy, with participants' autonomous motivation acting as a mediating variable. Furthermore, participants' increased self-efficacy was related to improved adherence to the dietary aspects of the treatment regimen. Consequently, training in the importance of autonomy supportive relationship styles should be undertaken within CF services to enable providers to offer optimal support to young people with CF. Further work, with a larger sample, is required to confirm that these factors are influential in increasing adherence to regimens within this population since this will improve the health of CF patients in the long-term and reduce the cost to the National Health Service.

CHAPTER 5: REFERENCES

- Andersen, D.H. (1938). Cystic fibrosis of the pancreas and its relation to coeliac diseases. A clinical and pathological study. *American Journal of Diseases in Childhood*. 56, 344-399.
- Anderson, R.M., Nowacek, G. & Richards, F. (1988). Influencing the personal meaning of diabetes: Research and practice. *Diabetes Educator*. 14, 297-302.
- Angst, D.B. (2001). School-age children. In Bluebond-Langer, M., Lask, B. & Angst, D.B. (Eds). *Psycho-social Aspects of Cystic Fibrosis*. pp.125-138. London. Arnold Publishers.
- Aspin, A.J. (1991). Psychological consequences of cystic fibrosis in adults. *Journal of Hospital Medicine*. 45, 368-371.
- Bandura, A. (1986). *Social Foundations of Thought and Action: A Social Cognitive Theory*. Englewood Cliffs. New Jersey. Prentice-Hall.
- Bandura, A. (1997). *Self-efficacy: The Exercise of Control*. New York. Freeman.
- Bartholomew, L.K., Parcel, G.S., Swank, P.R. & Czyzewski, D.I. (1993). Measuring self-efficacy expectations for the self-management of cystic fibrosis. *Chest*. 103(5), 1524-1530.
- Blair, C., Cull, A. & Freeman, C.P. (1994). Psycho-social functioning in young adults with cystic fibrosis and their families. *Thorax*. 49, 798-802.

- Bond, W.S. & Hussar, D.A. (1991). Detection methods and strategies for improving medication compliance. *American Journal of Hospital Pharmacy*. 48, 1978-1988.
- Bond, G.G., Aiken, L.S. & Somerville, S.C. (1992). The health belief model and adolescents with insulin-dependent diabetes mellitus. *Health Psychology*. 11, 190-198.
- Brown, C., Rowley, S. & Helms, P. (1994). Symptoms, health and illness behaviour in cystic fibrosis. *Social Science and Medicine*. 39, 375-379.
- Brugman, S.M. & Tausig, L.M. (1984). The reproductive system. In Tausig, L.M. (Ed). *Cystic Fibrosis*. pp.323-337. New York. Thieme-Stratton.
- Bush, A. (2001). Cystic fibrosis: Cause, course and treatment. In Bluebond-Langer, M., Lask, B. & Angst, D.B. (Eds). *Psycho-social Aspects of Cystic Fibrosis*. pp.1-26. London. Arnold Publishers.
- Cameron, K. & Gregor, F. (1987). Chronic illness and compliance. *Journal of Advanced Nursing*. 12, 671-676.
- Cappeli, M., McGrath, P.J., Heick, C.E., MacDonald, N.E., Feldman, W. & Rowe, P. (1987). Chronic disease and its impact. *Journal of Adolescent Health Care*. 10, 283-288.

Carstairs, V. & Morris, R. (1991). *Deprivation and Health in Scotland*. Aberdeen. Aberdeen University Press.

Christian, B.J. & D'Auria, J.P. (1997). The child's eye: Memories of growing up with cystic fibrosis. *Journal of Paediatric Nursing*. 12, 3-12.

Chumbley, J. (1999). *Cystic Fibrosis: A Family Affair. Overcoming Common Problems*. London. Sheldon Press.

Clark, N.M., Nothwehr, F., Gong, M., Evans, D., Maiman, L.A., Hurwitz, M.E., Roloff, D. & Mellins, R.B. (1995). Physician-patient partnership in managing chronic illness. *Academic Medicine*. Nov, 70(11), 957-958.

Cohen, J. (1992). Quantitative methods in psychology. A power primer. *Psychological Bulletin*. 112, 115-159.

Cohen, A. (1999). More trouble with ethics committees. *The British Journal of Psychiatry*. Oct, 175, 393.

Conway, S.P. (1998). Transition from paediatric to adult-orientated care for adolescents with cystic fibrosis. *Disability Rehabilitation*. June-July, 20(6-7), 209-216.

Conway, S.P., Pond, M.N., Hamnett, T. & Watson, A. (1996). Compliance with treatment in adult patients with cystic fibrosis. *Thorax*. 51, 29-33.

- Corey, M., McLaughlin, F.J., Williams, M. & Levison, H. (1988). A comparison of survival, growth and pulmonary function in patients with cystic fibrosis in Boston and Toronto. *Journal of Clinical Epidemiology*. 41, 588-591.
- Cowen, L., Coney, M., Simmons, R., Keenan, N., Robertson, J. & Levison, H. (1984). Growing older with cystic fibrosis: Psychological adjustment of patients more than 16 years old. *Psychosomatic Medicine*. 46, 363-376.
- Cuthbert, A.W. (1996). The cystic fibrosis gene. In Shale, D.J. (Ed). *Cystic Fibrosis*. pp.52-61. BMJ Publishing Group.
- Cystic Fibrosis Foundation. (1999). *Cystic Fibrosis Patient Registry 1998 Annual Data Report*. Bethesda, MD. Cystic Fibrosis Foundation.
- Czajkowski, D.R. & Koocher, G.P. (1987). Medical compliance and coping with cystic fibrosis. *Journal of Child Psychology and Psychiatry*. 28, 311-319.
- D'Angelo, S.L. & Lask, B. (2001). Approaches to problems of adherence. In Bluebond-Langer, M., Lask, B. & Angst, D.B. (Eds). *Psycho-social Aspects of Cystic Fibrosis*. pp.361-379. London. Arnold Publishers.
- Dark, J. & Corris, P. (1996). Transplantation. In Shale, D.J. (Ed). *Cystic Fibrosis*. pp.120-133. BMJ Publishing Group.

Deci, E.L. & Ryan, R.M. (1985). *Intrinsic Motivation and Self-Determination in Human Behaviour*. New York. Plenum Publishers.

Deci, E.L., Eghrari, H., Patrick, B.C. & Leone, D.R. (1994). Facilitating internalisation: The self-determination theory perspective. *Journal of Personality*. 62, 119-142.

Desmond, K.J. Schwenk, F., Thomas, E., Beaudry, P.H. & Coates, A.L. (1983). Immediate and long-term effects of chest physiotherapy in patients with cystic fibrosis. *Journal of Paediatrics*. 193, 538-542.

DiMatteo, M.R. & DiNicola, D.D. (1982). *Achieving Patient Compliance: The Psychology of the Medical Practitioner's Role*. New York. Pergamon Press.

Dunbar, J.M. & Agras, W.S. (1980). Compliance with medical instructions. In Ferguson, J.M. & Taylor, C.B. (Eds). *The Comprehensive Handbook of Behavioural Medicine. Volume 3*. pp.115-145. City Publisher.

Duncan-Skingle, F. & Pankhurst, F.J. (2001). Adults. In Bluebond-Langer, M., Lask, B. & Angst, D.B. (Eds). *Psycho-social Aspects of Cystic Fibrosis*. pp.161-171. London. Arnold Publishers.

Edenborough, F.P., Stableforth, D.E., Webb, A.K., Mackenzie, W.E. & Smith, D.L. (1995). Outcome of pregnancy in women with cystic fibrosis. *Thorax*. 50, 170-174.

Eiser, C. (1993). *Growing up with a Chronic Disease: The Impact on Children and their Families*. London. Jessica Kingsley Publishers.

Elborn, J.S., Shale, D.J. & Britton, J.R. (1991). Cystic fibrosis: Current survival and population estimates to the year 2000. *Thorax*. 46, 881-885.

Fakhoury, K., Durie, P.R., Levison, H. & Canny, G.J. (1992). Meconium ileus in the absence of fibrosis. *Archives of Diseases in Childhood*. 67, 1204-1206.

Felton, B.J., Revenson, T.A. & Hinrichsen, G.A. (1984). Stress and coping in the explanation of psychological adjustment among chronically ill adults. *Social Science and Medicine*. 18(10), 889-898.

FitzSimmons, S.C. (1993). The changing epidemiology of cystic fibrosis. *Journal of Paediatrics*. 122, 1-9.

Fowler, M.G., Johnson, M.P. & Atkinson, S.S. (1985). School achievement and absence in children with chronic health conditions. *Journal of Paediatrics*. 106, 683-687.

Gilbert, P. (2000). *Overcoming Depression: A Self-help Guide to Using Cognitive Behavioural Techniques*. London. Constable and Robinson Ltd.

Glaser, B.G. & Strauss, A.L. (1967). *The Discovery of Grounded Theory. Strategies for Qualitative Research*. New York. Aldine.

- Glasgow, R.E., McCaul, K.D. & Schafer, L.C. (1986). Barriers to regimen adherence among persons with insulin-dependent diabetes. *Journal of Behavioural Medicine*. 9, 65-77.
- Gotz, I. & Gotz, M. (2001). Reproduction and parenting. In Bluebond-Langer, M., Lask, B. & Angst, D.B. (Eds). *Psycho-social Aspects of Cystic Fibrosis*. pp.172-188. London. Arnold Publishers.
- Gudas, L.J., Koocher, G.P. & Wyplj, D. (1991). Perceptions of medical compliance in children and adolescents with cystic fibrosis. *Developmental and Behavioural Paediatrics*. 12, 236-247.
- Hains, A.A., Davies, W.H., Behrens, D. & Biller, J.A. (1997). Cognitive Behavioural Interventions for Adolescents with Cystic Fibrosis. *Journal of Paediatric Psychology*. 22, 669-687.
- Haynes, R.B. (1979). Introduction. In Haynes, R.B., Taylor, D.W. & Sackett, D.C. (Eds). *Compliance in Health Care*. pp.1-7. Baltimore. The John Hopkins University Press.
- Helton, J.L., Harmon, R.J. & Robinson, N. (1991). Parental attitudes towards newborn screening for CF. *Paediatric Pulmonary Supplement*. 7, 23-28.
- Hill, C.M. (1998). Diagnosis. In Hill, C.M. (Ed). *Practical Guidelines for Cystic Fibrosis Care*. pp.13-20. Churchill Livingstone.

Howells, L., Greene, A., Neville, R., Pagliari, C., Mukhopadhyay, S., Alexander, V. & Greene, S.A. (2001). Empowering young people for asthma self-management in a primary care setting. Submitted to the *Journal of Asthma*.

Jacobson, A.M., Hauser, S.T., Wolfsdorf, J.I., Houlihan, J., Milley, J.E., Herskowitz, R.D., Wertlieb, D. & Watt, E. (1987). Psychological predictors of compliance in children with recent onset diabetes mellitus. *Journal of Paediatrics*. 110, 805-811.

Koocher, G.D., McGrath, M.L. & Gudas, L.J. (1990). Typologies of non-adherence in cystic fibrosis. *Journal of Developmental and Behavioural Paediatrics*. 11, 353-358.

Kovacs, M., Goldston, D., Obrosky, D.S. & Iyengar, S. (1992). Prevalence and predictors of pervasive noncompliance with medical treatments among youths with insulin-dependent diabetes mellitus. *Journal of the American Academy of Child and Adolescent Psychiatry*. 31, 1112-1119.

Kyngas, H. & Barlow, J. (1995). Diabetes: An adolescent's perspective. *Journal of Advanced Nursing*. Nov, 22(5), 941-947.

Laine, C. & Davidoff, F. (1996). Patient-centered medicine: A professional evolution. *JAMA*. 27(5), 152-156.

Lask, B. (1994). Non-adherence in cystic fibrosis: Methods, meanings and management. *Journal of the Royal Society of Medicine*. 21, 25-27.

- Lawer, R.H., Nakielny, W. & Wright, N.A. (1996). Psychological implications of cystic fibrosis. *Canadian Medical Association Journal*. 94, 1043-1046.
- Lebow, J. (1982). Consumer satisfaction with mental health treatment. *Psychological Bulletin*. 119, 224-259.
- Lewin, L.O., Byard, P.J. & Davis, P.B. (1990). Effect of pseudomonas cepacia colonization on survival and pulmonary function of cystic fibrosis patients. *Journal of Clinical Epidemiology*. 43, 125-131.
- LiPuma, J.J., Dasen, S.E., Nielson, D.W., Stern, R.C. & Stull, T.L. (1990). Person to person transmission of pseudomonas cepacia between patients with cystic fibrosis. *Lancet*. 336, 1094-1097.
- Lorenz, R.A., Christensen, N.K. & Pichert, J.W. (1985). Diet-related knowledge, skill and adherence among children with insulin-dependent diabetes mellitus. *Paediatrics*. 75, 872-876.
- Masek, B. & Jankel, W.R. (1982). Therapeutic adherence. In Russo, D.C. & Varni, J.W. (Eds). *Behavioural Paediatrics*. pp.375-395. New York. Plenum Publishers.
- Moorey, S., Greer, S., Watson, M., Gorman, C., Rowden, L., Tunmore, R., Robertson, B. & Bliss, J. (1991). The factor structure and factor stability of the Hospital Anxiety and Depression Scale in patients with cancer. *British Journal of Psychiatry*. 158, 255-259.

- Mullins, L.L., Pace, T.M. & Keller, J. (1994). Cystic fibrosis: Psychological issues. In Olson, R.A., Mullins, L.L. & Chaney, J.M. (Eds). *The Sourcebook of Paediatric Psychology*. pp. 204-217. Boston. Allyn and Bacon.
- Nasr, S.Z. (2000). Cystic fibrosis in adolescents and young adults. *Adolescent Medicine*. Oct, 11(3), 589-603.
- O'Leary, A. (1992). Self-efficacy and health: Behavioural and stress-physiological mediation. *Cognitive Therapy and Research*. 16, 229-245.
- Parcel, G.S., Swank, P.R., Mariotto, M.J., Bartholomew, L.K., Czyzewski, D.I., Sockrider, M.M. & Seilheimer, D.K. (1994). Self-management of cystic fibrosis: A structural model for educational and behavioural variables. *Social Science and Medicine*. 38(9), 1307-1315.
- Passero, M.A., Remor, B. & Salomon, J. (1981). Patient-reported compliance with cystic fibrosis therapy. *Clinical Paediatrics*. 20, 264-268.
- PatientWise. (1997). *Medical and Health Information for Patients: Cystic Fibrosis*. No. 17.13. John Wiley & Sons Ltd.
- Patterson, J.M. (1985). Critical factors affecting family compliance with home treatment for children with cystic fibrosis. *Family Relations*. 34, 79-89.

Penketh, A.R., Wise, A., Mearns, M.B., Hodson, M.E. & Balten, J.C. (1987). Cystic fibrosis in adolescents and adults. *Thorax*, 42, 526-532.

Pownceby, J. (1997). *Coming of Age Project*. Cystic Fibrosis Trust. Kent.

Ramsey, B.W. & Marshall, S.G. (1995). Aerosol therapy in cystic fibrosis: DNase Tobramycin. *Seminars in Respiratory and Critical Care Medicine*. 15(5) 434-438.

Rapoff, M.A. (1999). *Adherence to Paediatric Medical Regimens*. Clinical Child Psychology Library. New York. Kluwer Academic. Plenum Publishers.

Rapoff, M.A. & Barnard, M.U. (1991). Compliance with paediatric medical regimens. In Cramer, J.A. & Spilker, B. (Eds). *Patient Compliance in Medical Practice and Clinical Trials*. pp.73-98. New York. Raven Press.

Reiter, E.O., Stern, R.C. & Root, A.W. (1981). The reproductive endocrine system in cystic fibrosis. 1. Basal gonadotrophin and sex steroid levels. *American Journal of Diseases in Childhood*. 135, 422-426.

Ricker, J.H., Delamater, A.M. & Hsu, J. (1998). Correlates of regimen adherence in cystic fibrosis. *Journal of Clinical Psychology in Medical Settings*. 5(2), 159-172.

Rosen, D. (1995). Between two worlds: Bridging the cultures of child health and adult medicine. *Journal of Adolescent Health*. 17, 10-16.

- Roy, C.C. (1983). Gastro-intestinal and hepatobiliary complications: Changing patterns with age. In Sturgess, J. (Ed). *Perspectives in Cystic Fibrosis*. pp.197. Ontario. Imperial Press.
- Rudd, P. (1993). The measurement of compliance: Medicine taking. In Krasnegor, M.A., Epstein, L., Johnson, S.B. & Yaffe, S.J. (Eds). *Developmental Aspects of Health Compliance Behaviour*. pp.185-213. New Jersey. Hillsdale. Erlbaum.
- Ryan, R.M. & Connell, J.P. (1989). Perceived locus of causality and internalisation: Examining reasons for acting in two domains. *Journal of Personality and Social Psychology*. 57, 749-761.
- Sensky, T. (1990). Patients' reactions to illness. *British Medical Journal*. 300, 622-623.
- Shale, D.J. (1996). Management in adults. In Shale, D.J. (Ed). *Cystic Fibrosis*. pp.14-31. BMJ Publishing Group.
- Shepherd, S., Hovell, M. & Harwood, I. (1990). A comparative study of the psychosocial assets of adults with cystic fibrosis and their healthy peers. *Chest*. 97, 1310-1316.
- Shwachman, H., Mahmoodian, A. & Neff, R.K. (1981). The sweat test: Sodium and chloride values. *Journal of Paediatrics*. 98, 576-578.

- Smith, D.L., Smith, E.G. & Gumery, L.B. (1992). *Pseudomonas cepacia* infection in cystic fibrosis. *Lancet*. 339-252.
- Strauss, G. & Wellisch, D. (1981). Psycho-social adaptation in older cystic fibrosis patients. *Journal of Chronic Diseases*. 34, 141-146.
- Suls, J. & Fletcher, B. (1985). The relative efficacy of avoidant and non-avoidant coping strategies: A meta-analysis. *Health Psychology*. 4, 249-288.
- Szaff, M., Hoiby, N. & Flensburg, E.W. (1983). Frequent antibiotic therapy improves survival of cystic fibrosis with chronic *pseudomonas aeruginosa* infection. *Acta Paediatrica Scandinavica*. 72, 651-657.
- Thompson, R.J., Hodge, S. K. & Hamlett, K.W. (1990). A matched comparison of adjustment in children with cystic fibrosis and psychiatrically referred and non-referred children. *Journal of Paediatric Psychology*. 15, 745-759.
- Tyrrell, J. (2001). Growing up with cystic fibrosis: The adolescent years. In Bluebond-Langer, M., Lask, B. & Angst, D.B. (Eds). *Psycho-social Aspects of Cystic Fibrosis*. pp.139-149. London. Arnold Publishers.
- Valerius, N.H., Koch, C. & Hoiby, N. (1991). Prevention of chronic *pseudomonas aeruginosa* colonisation in cystic fibrosis by early treatment. *Lancet*. 338, 725-726.

- Walters, S., Britton, J. & Hodson, M.E. (1993). Demographic and social characteristics of adults with cystic fibrosis in the United Kingdom. *British Medical Journal*. Feb, 306, 549-552.
- Warwick, W.J. (1982). Prognosis for survival with cystic fibrosis: Effects of early diagnosis and cystic fibrosis centre care. *Acta Paediatric Scandinavia Supplement*. 301, 27-31.
- Westwood, A., Henley, L. & Willcox, P. (1999). Transition from paediatric to adult care for persons with cystic fibrosis: Patient and parent perspectives. *Journal of Paediatrics and Child Health*. Oct, 35(5), 442-445.
- Williams, G.C. & Deci, E.L. (1996). The national cancer institute guidelines for smoking cessation: Do they motivate quitting?. *Journal of General International Medicine*. 11(1), 138.
- Williams, G.C. & Deci, E.L. (2001). Activating patients for smoking cessation through physician autonomy support. *Medical Care*. August, 39(8), 813-823.
- Williams, G.C., Frankel, R.M., Campbell, T.L. & Deci, E.L. (2000). Research on relationship-centred care and health care outcomes from the Rochester bio-psychosocial program: A self-determination theory integration. *Families, Systems and Health*. Spring, 18(1), 79-90.

Williams, G.C., Freedman, Z.R. & Deci, E.L. (1998a). Supporting autonomy to motivate patients with diabetes for glucose control. *Diabetes Care*. Oct, 21(10), 1644-1651.

Williams, G.C., Grow, V.M., Freedman, Z.R., Ryan, R.M. & Deci, E.L. (1996). Motivational predictors of weight-loss and weight-loss maintenance. *Journal of Personality and Social Psychology*. Jan, 70(1), 115-126.

Williams, G.C., Rodin, G.C., Ryan, R.M., Grolnick, W.S. & Deci, E.L. (1998b). Autonomous regulation and long-term medication adherence in adult outpatients. *Health Psychology*. May, 17(3), 269-276.

Williams, G.C., Saizow, R.B. & Ryan, R.M. (1999). The importance of self-determination theory for medical education. *Academic Medicine*. Sept, 74(9), 992-995.

Zigmond, A.S. & Snaith, R.P. (1983). The Hospital Anxiety and Depression Scale. *Acta Psychiatrica Scandinavica*. 67, 361-370.

APPENDIX 1

Questionnaire Pack

- (1) The Health Care Climate Questionnaire (HCCQ)
- (2) The Self-efficacy Questionnaire (SEQ)
- (3) The Treatment Self-regulation Questionnaire (TSRQ)
- (4) The Hospital Anxiety and Depression Scale (HADS)
- (5) A Self-report Measure of Adherence

Questionnaire Pack

Please answer the following questions and then complete the questionnaires below. There are no right or wrong answers and your responses will be kept private.

- A) Are you Male or Female ?
- B) How old are you ?
- C) Which care provider do you see most often ? (please tick)
CF doctor CF nurse..... Both seen as often

1. What do you think about the people who help with your Cystic Fibrosis?

These questions ask about how your care provider (e.g. CF doctor or nurse) deals with you and your CF. Care providers have different styles in dealing with CF patients. We would like to know more about how you feel about your meetings with your care provider. Put a number in the box to show how much you agree or not with the following statements.

1 =Strongly disagree
2 =Moderately disagree
3 =Slightly disagree
4 =Neutral

5 =Slightly agree
6 = Moderately agree
7 = Strongly agree

- | | |
|---|--|
| 1. <input type="checkbox"/> I feel that my care provider has provided me with choices and options. | 9. <input type="checkbox"/> My care provider answers my questions fully and carefully. |
| 2. <input type="checkbox"/> I feel understood by my care provider. | 10. <input type="checkbox"/> My care provider listens to how I would like to do things. |
| 3. <input type="checkbox"/> I am able to be open with my care provider at our meetings. | 11. <input type="checkbox"/> My care provider handles my emotions very well. |
| 4. <input type="checkbox"/> My care provider conveys confidence in my ability to make changes. | 12. <input type="checkbox"/> I feel that my care provider cares about me as a person. |
| 5. <input type="checkbox"/> I feel that my care provider accepts me (regardless of how well I look after my CF). | 13. <input type="checkbox"/> I don't feel very good about the way my care provider speaks to me. |
| 6. <input type="checkbox"/> My care provider has made sure that I really understand the risks of my condition and what I need to do, without pressuring me. | 14. <input type="checkbox"/> My care provider tries to understand how I see things before suggesting a new way to do things. |
| 7. <input type="checkbox"/> My care provider encourages me to ask questions. | 15. <input type="checkbox"/> I am able to share my feelings with my care provider. |
| 8. <input type="checkbox"/> I feel a lot of trust in my care provider. | |

2. How do you feel about your Cystic Fibrosis?

Put a number in the box to show how much you agree or not with the statements

1 =Strongly disagree
2 =Moderately disagree
3 =Slightly disagree
4 =Neutral

5 =Slightly agree
6 = Moderately agree
7 = Strongly agree

1. ☐ I feel confident in my ability to look after my CF.
2. ☐ I feel capable of handling my CF at the moment.
3. ☐ I am able to do my own routine CF care at the moment.
4. ☐ I am able to meet the challenge of looking after my CF.

3. Why do you look after your Cystic Fibrosis?

Think about the reasons why you look after your CF. Put a number in the box to show how much you agree or not with the following statements.

1 =Strongly disagree
2 =Moderately disagree
3 =Slightly disagree
4 =Neutral

5 =Slightly agree
6 = Moderately agree
7 = Strongly agree

1. ☐ Other people would be mad at me if I didn't.
2. ☐ I find it a personal challenge to keep my CF well controlled.
3. ☐ I believe that tightly controlling my CF will greatly improve my health.
4. ☐ I would feel guilty if I didn't do what my doctor said.
5. ☐ I believe looking after my CF is the most important reason for me remaining healthy.
6. ☐ Mostly, I want my doctor to think I'm a good patient.
7. ☐ Effectively looking after my CF is one of many important aspects of my life.
8. ☐ I would feel bad about myself if I didn't.
9. ☐ I feel personally satisfied when I keep my CF within strict guidelines.
10. ☐ It's rewarding to try to keep my lung function readings in a healthy range.
11. ☐ Managing my CF closely allows me to participate in other important aspects of my life.
12. ☐ I don't want other people to be disappointed in me.
13. ☐ Taking my medication (tablets and/or nebulizers) and doing my physiotherapy are a regular part of my life.

4. Read each item below and underline the reply which comes closest to how you have been feeling in the past week. Don't take too long over your replies, your immediate reaction to each item will probably be more accurate than a long thought-out response.

1. I feel tense or 'wound up'

Most of the time
A lot of the time
From time to time, occasionally
Not at all

2. I still enjoy the things I used to enjoy

Definitely as much
Not quite so much
Only a little
Hardly at all

3. I get a sort of frightened feeling as if something awful is about to happen

Very definitely and quite badly
Yes, but not too badly
A little, but it doesn't worry me
Not at all

4. I can laugh and see the funny side of things

As much as I always could
Not quite so much now
Definitely not so much now
Not at all

5. Worrying thoughts go through my mind

A great deal of the time
A lot of the time
Not too often
Very little

6. I feel cheerful

Never
Not often
Sometimes
Most of the time

7. I can sit at ease and feel relaxed

Definitely
Usually
Not often
Not at all

8. I feel as if I am slowed down

Nearly all the time
Very often
Sometimes
Not at all

9. I get a sort of frightened feeling like 'butterflies' in the stomach

Not at all
Occasionally
Quite often
Very often

10. I have lost interest in my appearance

Definitely
I don't take as much care as I should
I may not take quite as much care
I take just as much care as ever

11. I feel restless as if I have to be on the move

Very much indeed
Quite a lot
Not very much
Not at all

12. I look forward with enjoyment to things

As much as I ever did
Rather less than I used to
Definitely less than I used to
Hardly at all

13. I get sudden feelings of panic

Very often indeed
Quite often
Not very often
Not at all

14. I can enjoy a good book or radio or television programme

Often
Sometimes
Not often
Very seldom

5. Do you look after your Cystic Fibrosis in the way you are advised?

Draw a cross on the line at the point that shows how much you followed the CF advice given to you by your care provider over the past week.

1) MEDICATION
(e.g. antibiotics)

Every day

I often miss
some days

Never / I frequently
miss days

4

3

2

1

0

2) PHYSIOTHERAPY

Every day

I often miss
some days

Never / I only do my
physiotherapy if ill

4

3

2

1

0

3) DIET
(e.g. enzymes
and vitamins)

Every meal/snack

Most meals/snacks

Never

4

3

2

1

0

4) EXERCISE

Every day

I often miss
some days

Never

4

3

2

1

0

Would you say that the past week was a 'typical' week in terms of how much you followed the CF advice given to you by your care provider ?

Yes / No (Please delete)

If no, please describe what happened during the past week:

MANY THANKS FOR YOUR HELP

APPENDIX 2

Sample Questions from the Interview

SAMPLE QUESTIONS FROM THE INTERVIEW

These are examples of the questions that were asked during the interviews. The questions were not asked in any particular order and were flexible according to the respondent's replies.

Background Information

Please could you start by telling me a bit about yourself ?

Who do you live with ?

How do you get on with your family ?

Where do/did you go to school ?

Are you working at the moment ? What do you do ?

Has having CF affected your schooling/work ? In what way ?

When did you find out you had CF ?

Emotional Well-being

How do you feel about having CF ?

What effect has CF had whilst you were growing up (on social life/relationships) ?

What do you find most useful in helping you deal with having CF ?

Has there ever been a time when you found it difficult to cope with having CF ?

Who have been the most helpful people to you over the years ? In what ways ?

Information regarding Treatment Regimens

What treatments do you require for your CF?

How confident do you feel about carrying out your treatments ?

Which treatments are you most/least confident about doing ? Please explain why.

Is there any additional information you would like about your treatments ?

Which treatments do you find easiest/most difficult to adhere to ? Please explain why.

What are some of the reasons why you might not carry out your treatments ?

Relationships with Health Care Providers

How would you describe your relationship with your care providers (CF doctor/nurse) ?

Have there ever been times when you have felt that you have been listened to/have been understood by your care provider ?

If so, What happened ?

How did you feel ?

Have there ever been times when you have felt that you have not been listened to/have not been understood by your care provider ?

If so, What happened ?

How did you feel ?

What did you do ?

What would have made the situation better ?

Do you think that the services you receive at the paediatric/adult CF clinics could be better in any way?

Concerns about the Future

What are your thoughts about the future ?

Do you have any worries about your future health ?

Who do you talk to about these worries ?

Additional Questions

Do you have any questions that you would like to ask regarding this interview or the questionnaires that you completed ?

APPENDIX 3

Demographic Information Sheet

DEMOGRAPHIC INFORMATION SHEET

Name:

Age (in years):

Age at diagnosis (in months):

Where living:

Parental home
With partner/spouse
Alone
Other (describe)

Postcode:

Occupation:

Attends school
Higher education
Part-time work (describe)
Full-time work (describe)
Unemployed
Other (describe)

FEV1 % predicted (average reading for the previous six months):

Prescribed treatment regimen for CF:

Any other relevant information:
(e.g. sibling with CF, additional health problems)

APPENDIX 4

Introductory Letter to Participants in Tayside and Forth Valley

Clinical Psychology Department
Level 6, South Block
Ninewells Hospital & Medical School
Dundee. DD1 9SY

April 2002

Dear [patient's name]

Re: Invitation to participate in a study looking at the relationship between young people with cystic fibrosis and their health care providers and self-reports of adherence to cystic fibrosis treatment regimens.

I am carrying out a study at Ninewells Hospital, Dundee with young people who have cystic fibrosis. The study is being undertaken with Dr. Mukhopadhyay and Dr. Winter's knowledge and support. You are invited to take part in this study. I have enclosed an information sheet about the study, a consent form, a questionnaire pack and a pre-paid envelope. If you would like more information about the study or you have any questions then please do not hesitate to contact me at the Clinical Psychology Department on (01382) 425612. If you decide that you would like to participate in the study then please complete the consent form and questionnaire pack and return them in the pre-paid envelope within the next two weeks.

Thank You

Jackie Squire
Clinical Psychologist in Training with
Dr. Lesley Howells
Chartered Clinical Psychologist

Clinical Psychology Department
Level 6, South Block
Ninewells Hospital & Medical School
Dundee. DD1 9SY

May 2002

Dear [patient's name]

Re: Invitation to participate in a study looking at the relationship between young people with cystic fibrosis and their health care providers and self-reports of adherence to cystic fibrosis treatment regimens.

I am carrying out a study at Stirling Royal Infirmary and Falkirk and District Royal Infirmary with young people who have cystic fibrosis. The study is being undertaken with Dr. MacFadyen's knowledge and support. You are invited to take part in the study. I have enclosed an information sheet about the study, a consent form, a questionnaire pack and a pre-paid envelope. If you would like more information about the study or you have any questions then please do not hesitate to contact me at the Clinical Psychology Department on (01382) 425612. If you decide that you would like to participate in this study then please complete the consent form and questionnaire pack and return them in the pre-paid envelope within the next two weeks.

Thank You

Jackie Squire
Clinical Psychologist in Training with
Dr. Lesley Howells
Chartered Clinical Psychologist

APPENDIX 5

Information Sheet for Participants in Tayside and Forth Valley

Research Information Sheet for Participants

I am carrying out a project with young people who have cystic fibrosis. The study is being undertaken with Dr. Mukhopadhyay and Dr. Winter's knowledge and support. You are invited to take part in the project that will look at factors that may be important to young people to encourage them to adhere to their treatment. This information sheet has been developed to help you to understand what the project is about so that you can decide whether or not you want to take part.

What is the research project all about ?

I am interested in your views about meeting with your care provider (e.g. the doctor/nurse at the Cystic Fibrosis Clinic) and what your care provider has told you about your condition and its treatment. I am also interested in your thoughts and feelings about having cystic fibrosis and how well you follow the advice from your care provider and look after yourself. I hope that the information gathered will allow me to provide better information to care providers about how to work with young people to encourage them to adhere to their treatment.

What will I have to do ?

If you agree to take part in the study, you will be asked to fill out five short questionnaires that will ask about your views on cystic fibrosis and how it affects your daily life. The questionnaires will take approximately 30 minutes to complete in total. There are no right or wrong answers to the questions. Not all of the questions in the questionnaires may apply to you and the questions that may not apply to you do not necessarily have to be answered. Your responses to the questionnaires are confidential and your answers will not be shared with your care provider or your parents/guardian. If you appear to be having difficulties in coping with your condition, your care provider may be informed of this but only with your permission. I will also need access to parts of your medical file to gather further information about the treatments offered to you.

It is your choice whether or not you want to take part in this project. If you agree to take part and then change your mind you can withdraw from the study, without giving a reason, and it will not affect your future medical care in anyway. If you do want to take part in the project then please sign the consent form and complete the questionnaire pack and return them both in the pre-paid envelope provided **within the next two weeks**. A copy of your consent form will be sent to your GP so they are aware that you are assisting me with the project.

In addition to completing the questionnaires, you are invited to attend an interview to discuss your experiences of having cystic fibrosis in some more detail. If you agree to take part in the interview, I will arrange to meet with you on one occasion at your home or Ninewells Hospital, Dundee, whichever is most convenient for you, for approximately 40 minutes. The information obtained during this meeting will be confidential. The interview will be tape recorded with your consent and the audio tapes will be destroyed at the end of the study.

What if I want any more information before making a decision ?

If you have any questions about this information sheet or you would like more information about the project then please do not hesitate to contact me at the Clinical Psychology Department, Ninewells Hospital on (01382) 425612.

Thank you for taking the time to read this information sheet.

Research Information Sheet for Participants

I am carrying out a project with young people who have cystic fibrosis. The study is being undertaken with Dr. MacFadyen's knowledge and support. You are invited to take part in the project that will look at factors that may be important to young people to encourage them to adhere to their treatment. This information sheet has been developed to help you to understand what the project is about so that you can decide whether or not you want to take part.

What is the research project all about ?

I am interested in your views about meeting with your care provider (e.g. the doctor/nurse at the Cystic Fibrosis Clinic) and what your care provider has told you about your condition and its treatment. I am also interested in your thoughts and feelings about having cystic fibrosis and how well you follow the advice from your care provider and look after yourself. I hope that the information gathered will allow me to provide better information to care providers about how to work with young people to encourage them to adhere to their treatment.

What will I have to do ?

If you agree to take part in the study, you will be asked to fill out five short questionnaires that will ask about your views on cystic fibrosis and how it affects your daily life. The questionnaires will take approximately 30 minutes to complete in total. There are no right or wrong answers to the questions. Not all of the questions in the questionnaires may apply to you and the questions that may not apply to you do not necessarily have to be answered. Your responses to the questionnaires are confidential and your answers will not be shared with your care provider or your parents/guardian. If you appear to be having difficulties in coping with your condition, your care provider may be informed of this but only with your permission. I will also need access to parts of your medical file to gather further information about the treatments offered to you.

It is your choice whether or not you want to take part in this project. If you agree to take part and then change your mind you can withdraw from the study, without giving a reason, and it will not affect your future medical care in anyway. If you do want to take part in the project then please sign the consent form and complete the questionnaire pack and return them both in the pre-paid envelope provided **within the next two weeks**. A copy of your consent form will be sent to your GP so they are aware that you are assisting me with the project.

In addition to completing the questionnaires, you are invited to attend an interview to discuss your experiences of having cystic fibrosis in some more detail. If you agree to take part in the interview, I will arrange to meet with you on one occasion at your home, Stirling Royal Infirmary or Falkirk and District Royal Infirmary, whichever is most convenient for you, for approximately 40 minutes. The information obtained during this meeting will be confidential. The interview will be tape recorded with your consent and the audio tapes will be destroyed at the end of the study.

What if I want any more information before making a decision ?

If you have any questions about this information sheet or you would like more information about the project then please do not hesitate to contact me at the Clinical Psychology Department, Ninewells Hospital on (01382) 425612.

Thank you for taking the time to read this information sheet.

APPENDIX 6

Consent Form

Consent Form

Title of Project:

The relationship between young people with cystic fibrosis and their health care providers and self-reports of adherence to cystic fibrosis treatment regimens.

Name of Researcher:

Jackie Squire (under the supervision of Lesley Howells)

Please Tick

- 1) I confirm that I have read and understood the information sheet for the above study and have had the opportunity to ask questions. ☐
- 2) I understand that my participation is voluntary and that I am free to withdraw at any time, without giving a reason, without my medical care or legal rights being affected. ☐
- 3) I understand that sections of my medical notes will be looked at by Jackie, Squire, Clinical Psychologist in Training, where it is relevant to my taking part in the research. I give permission for this individual to have access to my records and to use the information on the understanding that all identifying features will be removed to ensure anonymity. ☐
- 4) I agree to take part in the study. ☐
- 5) I agree to be interviewed and agree to the interview being tape recorded on the understanding that the tape is destroyed at the end of the study. ☐

Name of patient _____

Signature _____ **Date** _____

Signature of Parent (if under 16 years) _____ **Date** _____

Name of Researcher _____

Signature _____ **Date** _____

APPENDIX 7

Reply Slip for the Interview

REPLY SLIP FOR THE INTERVIEW

I would / would not be willing to be interviewed about my experiences of having cystic fibrosis **(Please delete)**

If you have decided to take part in the interview then please complete the following sections and return this form with the completed consent form and questionnaire pack in the pre-paid envelope provided **within the next two weeks**. If you do not wish to take part in the interview then you do not need to return this form.

Name:

Telephone number:

Best day and time for appointment **(Please tick)**

	Monday	Tuesday	Wednesday	Thursday	Friday	Any Day
a.m.						
p.m.						

Are there any days that would be unsuitable over the next month ?

Thank you.

Jackie Squire
(Clinical Psychologist in Training with Dr Lesley Howells, Clinical Psychologist).

APPENDIX 8

Letter sent to Participants' GPs

Clinical Psychology Department
Level 6, South Block
Ninewells Hospital & Medical School
Dundee. DD1 9SY

[Date]

Dear [GP's name]

Re: [Patient's name]

The above named individual has given their consent to participate in the following research project: **The relationship between young people with cystic fibrosis and their health care providers and self-reports of adherence to cystic fibrosis treatment regimens.**

The study will be undertaken at Ninewells Hospital, Dundee from April 2002 until July 2002. If you would like further information or have any questions about the research project then please do not hesitate to contact me at the Clinical Psychology Department on (01382) 425612.

I have enclosed a copy of [patient's name] signed consent form for your files.

Yours sincerely

Jackie Squire
Clinical Psychologist in Training with
Dr. Lesley Howells
Chartered Clinical Psychologist

Clinical Psychology Department
Level 6, South Block
Ninewells Hospital & Medical School
Dundee. DD1 9SY

[Date]

Dear [GP's name]

Re: [Patient's name]

The above named individual has given their consent to participate in the following research project: **The relationship between young people with cystic fibrosis and their health care providers and self-reports of adherence to cystic fibrosis treatment regimens.**

The study will be undertaken at Stirling Royal Infirmary and Falkirk and District Infirmary from May 2002 until July 2002. If you would like further information or have any questions about the research project then please do not hesitate to contact me at the Clinical Psychology Department on (01382) 425612.

I have enclosed a copy of [patient's name] signed consent form for your files.

Yours sincerely

Jackie Squire
Clinical Psychologist in Training with
Dr. Lesley Howells
Chartered Clinical Psychologist

APPENDIX 9

Introductory Letter to Parents in Tayside and Forth Valley

***This letter was sent to parents/guardians if their son/daughter was under 16 years of age.**

Clinical Psychology Department
Level 6, South Block
Ninewells Hospital & Medical School
Dundee. DD1 9SY

April 2002

Dear [parent/guardian]

Re: Invitation to participate in a study looking at the relationship between young people with cystic fibrosis and their health care providers and self-reports of adherence to cystic fibrosis treatment regimens.

I am carrying out a study at Ninewells Hospital, Dundee with young people who have cystic fibrosis. The study is being undertaken with Dr. Mukhopadhyay and Dr. Winter's knowledge and support. Your [son/daughter] is invited to take part in this study. I have enclosed an information sheet about the study for your perusal in addition to a consent form, a questionnaire pack and a pre-paid envelope. If you would like more information about the study or you have any questions then please do not hesitate to contact me at the Clinical Psychology Department on (01382) 425612. If you are happy for your [son/daughter] to participate in the study, please countersign the consent form and give the questionnaire pack to your [son/daughter] for completion. It would be appreciated if the completed consent form and questionnaire pack could be returned in the pre-paid envelope within the next two weeks.

Thank You

Jackie Squire
Clinical Psychologist in Training with
Dr. Lesley Howells
Chartered Clinical Psychologist

***This letter was sent to parents/guardians if their son/daughter was under 16 years of age.**

Clinical Psychology Department
Level 6, South Block
Ninewells Hospital & Medical School
Dundee. DD1 9SY

May 2002

Dear [parent/guardian]

Re: Invitation to participate in a study looking at the relationship between young people with cystic fibrosis and their health care providers and self-reports of adherence to cystic fibrosis treatment regimens.

I am carrying out a study at Stirling Royal Infirmary and Falkirk and District Royal Infirmary with young people who have cystic fibrosis. The study is being undertaken with Dr. MacFadyen's knowledge and support. Your [son/daughter] is invited to take part in this study. I have enclosed an information sheet about the study for your perusal in addition to a consent form, a questionnaire pack and a pre-paid envelope. If you would like more information about the study or you have any questions then please do not hesitate to contact me at the Clinical Psychology Department on (01382) 425612. If you are happy for your [son/daughter] to participate in the study, please countersign the consent form and give the questionnaire pack to your [son/daughter] for completion. It would be appreciated if the completed consent form and questionnaire pack could be returned in the pre-paid envelope within the next two weeks.

Thank You

Jackie Squire
Clinical Psychologist in Training with
Dr. Lesley Howells
Chartered Clinical Psychologist

APPENDIX 10

Information Sheet for Parents in Tayside and Forth Valley

Research Information Sheet for Parent/Guardian

I am carrying out a project with young people who have cystic fibrosis. The study is being undertaken with Dr. Mukhopadhyay and Dr. Winter's knowledge and support. Your [son/daughter] is invited to take part in the project that will look at factors that may be important to young people to encourage them to adhere to their treatment. This information sheet has been developed for your perusal to help you to understand what the project is about so that you can decide whether or not you are in agreement with your [son/daughter] taking part.

What is the research project all about ?

I am interested in the views of your [son/daughter] about meeting with their care provider (e.g. the doctor/nurse at the Cystic Fibrosis Clinic) and what their care provider has told them about their condition and its treatment. I am also interested in your [son/daughter]'s thoughts and feelings about having cystic fibrosis and how well they follow the advice from their care provider and look after themselves. I hope that the information gathered will enable me to provide better information to care providers about how to work with young people to encourage them to adhere to their treatment.

What will my [son/daughter] have to do ?

If you are in agreement with your [son/daughter] taking part in the study, they will be asked to complete five short questionnaires that will ask your [son/daughter] about their views on cystic fibrosis and how it affects their daily life. The questionnaires will take approximately 30 minutes to complete in total. Your [son/daughter]'s responses to the questionnaires are confidential and their answers will not be shared with their care provider or yourselves, in order to increase the likelihood that they will answer the questionnaires honestly. If your [son/daughter] appears to be having difficulties in coping with their condition, their care provider may be informed but only with your [son/daughter]'s permission. I will also need access to parts of your [son/daughter]'s medical file to gather further information about the treatments offered to them.

Your [son/daughter] can choose whether or not they want to take part in this project. If they agree to take part and then change their mind they can withdraw from the study, without having to give a reason, and it will not affect their future medical care in anyway. If your [son/daughter] wants to take part in the project and you are in agreement with this, then please can your [son/daughter] sign the consent form. As your

[son/daughter] is under 16 years of age, please could you also sign the consent form as their parent/guardian. The questionnaire pack should then be completed by your [son/daughter] and be returned with the signed consent form in the pre-paid envelope provided **within the next two weeks**. A copy of the consent form will be sent to your [son/daughter]'s GP so that their GP is aware that they are assisting me with the project.

In addition to completing the questionnaires, your [son/daughter] is invited to attend an interview to discuss their experiences of having cystic fibrosis in some more detail. If your [son/daughter] agrees to take part in the interview, I will arrange to meet with them on one occasion at their home or Ninewells Hospital, Dundee, whichever is most convenient, for approximately 40 minutes. The information obtained during this meeting will be confidential. The interview will be tape recorded with both you and your [son/daughter]'s consent and the audio tapes will be destroyed at the end of the study.

What if I want more information before making a decision ?

If you have any questions about this information sheet or you would like more information about the project then please do not hesitate to contact me at the Clinical Psychology Department, Ninewells Hospital on (01382) 425612.

Thank you for taking the time to read this information sheet.

Research Information Sheet for Parent/Guardian

I am carrying out a project with young people who have cystic fibrosis. The study is being undertaken with Dr. MacFadyen's knowledge and support. Your [son/daughter] is invited to take part in the project that will look at factors that may be important to young people to encourage them to adhere to their treatment. This information sheet has been developed for your perusal to help you to understand what the project is about so that you can decide whether or not you are in agreement with your [son/daughter] taking part.

What is the research project all about ?

I am interested in the views of your [son/daughter] about meeting with their care provider (e.g. the doctor/nurse at the Cystic Fibrosis Clinic) and what their care provider has told them about their condition and its treatment. I am also interested in your [son/daughter]'s thoughts and feelings about having cystic fibrosis and how well they follow the advice from their care provider and look after themselves. I hope that the information gathered will enable me to provide better information to care providers about how to work with young people to encourage them to adhere to their treatment.

What will my [son/daughter] have to do ?

If you are in agreement with your [son/daughter] taking part in the study, they will be asked to complete five short questionnaires that will ask your [son/daughter] about their views on cystic fibrosis and how it affects their daily life. The questionnaires will take approximately 30 minutes to complete in total. Your [son/daughter]'s responses to the questionnaires are confidential and their answers will not be shared with their care provider or yourselves, in order to increase the likelihood that they will answer the questionnaires honestly. If your [son/daughter] appears to be having difficulties in coping with their condition, their care provider may be informed but only with your [son/daughter]'s permission. I will also need access to parts of your [son/daughter]'s medical file to gather further information about the treatments offered to them.

Your [son/daughter] can choose whether or not they want to take part in this project. If they agree to take part and then change their mind they can withdraw from the study, without having to give a reason, and it will not affect their future medical care in anyway. If your [son/daughter] wants to take part in the project and you are in agreement with this, then please can your [son/daughter] sign the consent form. As your

[son/daughter] is under 16 years of age, please could you also sign the consent form as their parent/guardian. The questionnaire pack should then be completed by your [son/daughter] and be returned with the signed consent form in the pre-paid envelope provided **within the next two weeks**. A copy of the consent form will be sent to your [son/daughter]'s GP so that their GP is aware that they are assisting me with the project.

In addition to completing the questionnaires, your [son/daughter] is invited to attend an interview to discuss their experiences of having cystic fibrosis in some more detail. If your [son/daughter] agrees to take part in the interview, I will arrange to meet with them on one occasion at their home, Stirling Royal Infirmary or Falkirk and District Royal Infirmary, whichever is most convenient, for approximately 40 minutes. The information obtained during this meeting will be confidential. The interview will be tape recorded with both you and your [son/daughter]'s consent and the audio tapes will be destroyed at the end of the study.

What if I want more information before making a decision ?

If you have any questions about this information sheet or you would like more information about the project then please do not hesitate to contact me at the Clinical Psychology Department, Ninewells Hospital on (01382) 425612.

Thank you for taking the time to read this information sheet.

APPENDIX 11

Reminder Letter to Participants/Parents

May 2002

Dear [patient's name]

Re: Study looking at the relationship between young people with cystic fibrosis and their health care providers and self-reports of adherence to cystic fibrosis treatment regimens.

I wrote to you in April 2002 regarding a study that I am undertaking at Ninewells Hospital, Dundee with Dr Mukhopadhyay and Dr. Winter's knowledge and support. The study aims to look at factors that may be important to young people to encourage them to adhere to their treatment for cystic fibrosis. The first stage of the study will require you to complete five questionnaires that will take approximately 30 minutes in total. The second stage of the study will involve meeting with you for an interview lasting approximately 40 minutes to discuss your experiences of having cystic fibrosis in some more detail.

- If you do not wish to participate in the study or you have already returned the signed consent form and completed questionnaire pack in the pre-paid envelope then please ignore this reminder letter.
- If you have not yet returned the signed consent form and would like additional information about the study then please do not hesitate to contact me at the Clinical Psychology Department, Ninewells Hospital on (01382) 425612.
- If you are reluctant to meet with myself to discuss your experiences in detail but are happy to complete the questionnaires at home, please could you ensure that the signed consent form and completed questionnaire pack are returned in the pre-paid envelope provided **within the next two weeks**.

Thank you for your co-operation.

Jackie Squire

(Clinical Psychologist in Training with Dr. Lesley Howells, Clinical Psychologist).

***This letter was sent to parents/guardians if their son/daughter was under 16 years of age.**

Clinical Psychology Department
Level 6, South Block
Ninewells Hospital & Medical School
Dundee. DD1 9SY

May 2002

Dear [parent/guardian]

Re: Study looking at the relationship between young people with cystic fibrosis and their health care providers and self-reports of adherence to cystic fibrosis treatment regimens.

I wrote to you in April 2002 regarding a study that I am undertaking at Ninewells Hospital, Dundee with Dr. Mukhopadyay and Dr. Winter's knowledge and support. The study aims to look at factors that may be important to young people to encourage them to adhere to their treatment for cystic fibrosis. The first stage of the study will require your [son/daughter] to complete five questionnaires that will take approximately 30 minutes in total. The second stage of the study will involve meeting with your [son/daughter] for an interview lasting approximately 40 minutes to discuss their experiences of having cystic fibrosis in some more detail.

- If you do not wish your [son/daughter] to participate in the study or you have already returned the signed consent form and completed questionnaire pack in the pre-paid envelope then please ignore this reminder letter.
- If you have not yet returned the signed consent form and would like additional information about the study then please do not hesitate to contact me at the Clinical Psychology Department, Ninewells Hospital on (01382) 425612.
- If your [son/daughter] is reluctant to meet with myself to discuss their experiences in detail but are happy to complete the questionnaires at home, please could you ensure that the signed consent form and completed questionnaire pack are returned in the pre-paid envelope provided **within the next two weeks**.

Thank you for your co-operation.

Jackie Squire

(Clinical Psychologist in Training with Dr. Lesley Howells, Clinical Psychologist).

May 2002

Dear [patient's name]

Re: Study looking at the relationship between young people with cystic fibrosis and their health care providers and self-reports of adherence to cystic fibrosis treatment regimens.

I wrote to you at the beginning of May 2002 regarding a study that I am undertaking at Stirling Royal Infirmary and Falkirk and District Royal Infirmary, with Dr. MacFadyen's knowledge and support. The study aims to look at factors that may be important to young people to encourage them to adhere to their treatment for cystic fibrosis. The first stage of the study will require you to complete five questionnaires that will take approximately 30 minutes in total. The second stage of the study will involve meeting with you for an interview lasting approximately 40 minutes to discuss your experiences of having cystic fibrosis in some more detail.

- If you do not wish to participate in the study or you have already returned the signed consent form and completed questionnaire pack in the pre-paid envelope then please ignore this reminder letter.
- If you have not yet returned the signed consent form and would like additional information about the study then please do not hesitate to contact me at the Clinical Psychology Department, Ninewells Hospital on (01382) 425612.
- If you are reluctant to meet with myself to discuss your experiences in detail but are happy to complete the questionnaires at home, please could you ensure that the signed consent form and completed questionnaire pack are returned in the pre-paid envelope provided **within the next two weeks**.

Thank you for your co-operation.

Jackie Squire

(Clinical Psychologist in Training with Dr. Lesley Howells, Clinical Psychologist).

***This letter was sent to parents/guardians if their son/daughter was under 16 years of age.**

Clinical Psychology Department
Level 6, South Block
Ninewells Hospital & Medical School
Dundee. DD1 9SY

May 2002

Dear [parent/guardian]

Re: Study looking at the relationship between young people with cystic fibrosis and their health care providers and self-reports of adherence to cystic fibrosis treatment regimens.

I wrote to you at the beginning of May 2002 regarding a study that I am undertaking at Stirling Royal Infirmary and Falkirk and District Royal Infirmary, with Dr. MacFadyen's knowledge and support. The study aims to look at factors that may be important to young people to encourage them to adhere to their treatment for cystic fibrosis. The first stage of the study will require your [son/daughter] to complete five questionnaires that will take approximately 30 minutes in total. The second stage of the study will involve meeting with your [son/daughter] for an interview lasting approximately 40 minutes to discuss their experiences of having cystic fibrosis in some more detail.

- If you do not wish your [son/daughter] to participate in the study or you have already returned the signed consent form and completed questionnaire pack in the pre-paid envelope then please ignore this reminder letter.
- If you have not yet returned the signed consent form and would like additional information about the study then please do not hesitate to contact me at the Clinical Psychology Department, Ninewells Hospital on (01382) 425612.
- If your [son/daughter] is reluctant to meet with myself to discuss their experiences in detail but are happy to complete the questionnaires at home, please could you ensure that the signed consent form and completed questionnaire pack are returned in the pre-paid envelope provided **within the next two weeks.**

Thank you for your co-operation.

Jackie Squire

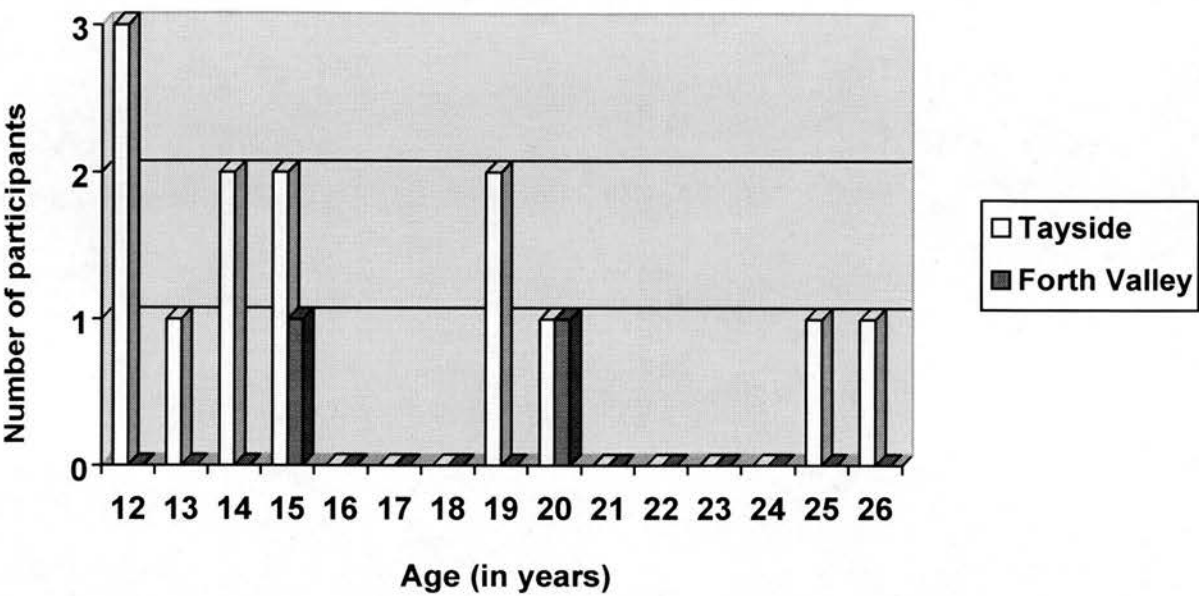
(Clinical Psychologist in Training with Dr. Lesley Howells, Clinical Psychologist).

APPENDIX 12

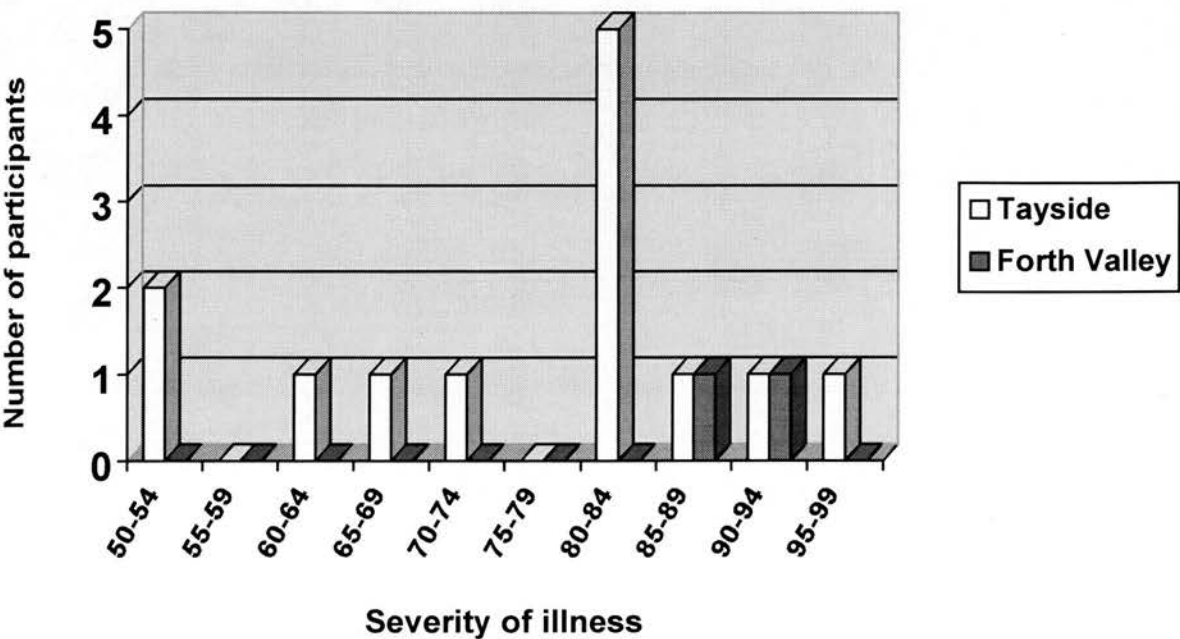
Graphs of Participants' Demographic Details

- (1) Age of Participants from Tayside and Forth Valley
- (2) Severity of Illness of Participants from Tayside and Forth Valley
- (3) Social Deprivation Categories of Participants from Tayside and Forth Valley

(1) Graph of age of participants from Tayside and Forth Valley (n=15)



(2) Graph of severity of illness of participants from Tayside and Forth Valley (n=15)
(as measured by FEV1 % predicted)



(3) Graph of social deprivation categories of participants from Tayside and Forth Valley (n=15)

